MINISTRY OF HEALTH OF UKRAINE
"Ukrainian Medical Dental Academy"

«Approved»
on meeting the
department of Pediatric Surgery
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The Head of the department
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METHODICAL INSTRUCTIONS

FOR STUDENTS` SELF-WORK

WHILE PREPARING FOR PRACTICAL LESSONS

Educational discipline	Pediatric Surgery
module №3	Urgent Pediatric Surgery
Theme of the lesson	Tumors of the skin and soft tissues. Neoplasms of bone. Nephroblastoma and Neuroblastoma. Tumors of the mediastinum.
Course	V
Faculty	foreign students preparation

POLTAVA 2020

1. The topic basis: the topic “Tumors of the skin and soft tissues. Neoplasms of bone. Nephroblastoma and Neuroblastoma. Tumors of the mediastinum.” is very important for future doctors in their professional activity, positively influences the students in their attitude to the future profession, forms professional skills and experience as well as taking as a principle the knowledge of the subject learnt.

2. The aims of the training course:

1. To master diseases which cause a formation of soft tissue.
2. To recognize the basic clinical manifestation of formations of soft tissue.
3. To differentiate new formation depending on their type.
4. To find out the signs of malignant regeneration of formations and their complications (bleeding, inflammations, squeezing of nerve and vessels etc).
5. To interpret principles of treatment of formations of soft tissue and their complications.
6. To recognize the basic clinical manifestation of tumours of bones, distinguish the signs of malignant regeneration.
7. To identify the features of course of separate diseases on the basis of clinical and X-ray signs.
8. To differentiate the innocent and malignant tumours of bones on the basis of clinical and auxiliary methods of research (X-ray, USD, CT, angiography, puncture and the opened biopsy of tumour).
9. To work out a plan of patient examination and algorithm of the actions of the doctor at complications of tumours of bones (pathological fractures, false joints, metastasis etc).
10. To interpret principles of surgical treatment, chemotherapy, radial therapy depending on the type of tumour of bone and the type of the course (innocent or malignant).
11. To master the characteristic signs of syndrome of "palpable tumour of abdomen" at nephroblastoma and neuroblastoma.
12. To distinguish nephroblastoma and neuroblastoma depending on clinical manifestation and stage of the course of diseases.
13. To interpret auxiliary methods of research: (USD, X-ray, urography, pneumoperitoneum, CT, puncture biopsy).
14. To conduct differential diagnostics of nephroblastoma, neuroblastoma, hydronephrosis, polycystic kidney, duplex kidney, tumours of liver, tumours of adrenal gland, lyphogenic tumours.
15. To explain principles of complex treatment of nephroblastoma and nephroblastoma depending on the stage of the course of the disease.
16. To learn the classification of innocent and malignant tumours of mediastinum widespread in children.
17. To recognize the basic clinical nanifestations of tumours of mediastinum.
18. To differentiate tumours of mediastinum.
19. To show the examination of a child with the tumors of mediastinum, to define its localization and boundaries.
20. To identify the features of the course of tumours of mediastinum.

3. Basic knowledge, skills, habits necessary for studying the subject (interdisciplinary integration).

Names of previous disciplines	Obtained skills

1. Anatomy	Describe the anatomy of the abdominal, thoracic cavity and musculoskeletal system. To assess the features of possible variants of the anatomical structure of the thoracic and abdominal cavities, to determine the features of the structure of bones in different age periods.
2. Hystology	Know the histological picture of the abdominal, thoracic cavity and musculoskeletal system. Be able to determine the features of the histological picture of different parts of the gastrointestinal tract, the chest and bone in children of different age groups
3. Operative surgery	To depict schematically the peculiarities of surgical interventions in children. To determine the features of topographic anatomy of the thoracic, abdominal cavities, musculoskeletal system in children of different age groups; To justify operative dissections and interventions depending on the pathology and age of the child
4.Propedeutics of childhood diseases	Possess the technique of examining a child with tumors of the thoracic, abdominal cavity, musculoskeletal system. To assess and demonstrate the knowledge of clinical and laboratory studies, the main symptoms of inflammatory diseases of the thoracic and abdominal organs, the main clinical symptoms are characteristic of diseases of the musculoskeletal system.
5.Patanatomy	Infectious pathologoanatomical changes in tumors and diseases of the respiratory system, gastrointestinal tract, musculoskeletal system. Identify specific pathological changes, their sequence for tumors of the thoracic, abdominal cavities in children of different ages and tumors of the musculoskeletal system.
6. Biochemistry	Demonstrate laboratory methods of examining a child with tumors of the abdominal, thoracic cavity and musculoskeletal system. To evaluate the data of clinical and biochemical analyzes: glucose in blood tests, urine; Protein in the blood serum, urine; Microelements
7. Pathological physiology	Describe pathological changes in tumors of diseases of the respiratory system, gastrointestinal tract, musculoskeletal system. To determine the main points of etiology, pathogenesis in acute surgical diseases, tumors of the thoracic, abdominal cavities in children of different ages.
8. physiology	Describe the physiology of the gastrointestinal tract, respiratory system, musculoskeletal system. Identify the features of the respiratory system and digestive system in

	a child of this age

Thereotical questions for the lesson:

1. Name the clinical manifestations of hemangiom and lymphangiom.
2. Name the methods of conservative and operative treatment of hemangiom and lymphangiom.
3. Name the clinical manifestations of pigmental tumours.
4. Medical tactic at pigmental tumours.
5. Clinic manifestations of atheroma and dermoid cysts.
6. Peculiarities of dermoid cysts removal.
7. Name the clinical manifestations of melanoma.
8. Differential diagnostics and combined treatment of melanoma.
9. Name the clinical manifestations of malignant tumours of soft tissue (rabdomyosarcoma, teratoblastoma).
10. Surgical and combined treatment of malignant tumours of soft tissue.
11. Auxiliary methods of diagnostics of tumours at children, and their role in differential diagnostics.
12. Peculiarities of clinical development of neoplasm and neoplasm-like lesion of bones.
13. Methods of surgical treatment of neoplasm and neoplasm-like lesion of bones.
14. Conservative treatment of cysts of bones.
15. Clinical manifestations of osteosarcoma and Ewing's sarcoma.
16. Principles of the combined treatment of malignant tumors of bones.
17. Clinical manifestations of innocent tumours of bones.
18. Methods of treatment of innocent tumours of bones.
19. Methods of diagnostics of malignant tumours of bones.
20. Clinical manifestations and treatment of nefroblstoma at children.
21. Clinical manifestations and treatment of neuroblastoma at children.
22. Auxiliary methods of diagnostics of tumours of retroperitoneum at children.
23. Clinic and diagnostics of tumours of the mediastinum.
24. Treatment of children with the malignant and innocent tumours of the mediastinum.

4. Maintenance of the subject:

TUMORS OF VASCULAR ORIGIN

These are the most common tumors of early life. Most of them are probably hamartomas rather than true neoplasms, and most are benign. They may occur in any organ, but there are sites of predilection: the skin and subcutaneous tissue , skeletal muscle, liver, salivary gland, larynx and bone.

Cavernous hemangiomas are poorly circumscribed, blue or purple elevated tumors which tend to extend more deeply into the subcutaneous tissues than do the capillary hemangiomas. They consist of numerous cystic vascular spaces containing blood. Mixed forms of cavernous and capillary hemangiomas are common; in such lesions regression of the cavernous elements may be slower and less constant than that of the superficial capillary elements. Cavernous elements which do not regress spontaneously are probably best treated by surgical excision. Cavernous hemangiomas may occur in sites other than the skin, e.g., in bone, liver and the tongue. In skeletal muscles they manifest themselves as a diffuse mass, sometimes accompanied by pain; the mass decreases in size with elevation of the part. Foci of calcification may be demonstrable by roentgenographic examination.

A giant hemangioma, usually of the skin or subcutaneous tissue, but infrequently a visceral one, may be accompanied by thrombocytopenia, bleeding and a consumptive coagulopathy. This is most often seen in the first year of life with a large, rapidly growing cavernous hemangioma, but

may occur at a later date in children with multiple hemangiomas that do not involute. Treatment, if necessary, has included the administration of corticosteroids, platelet transfusions, replacement of clotting factors, the use of anticoagulants and/or irradiation. Diffuse hemangiomas of an extremity may be associated with hypertrophy of the part and of the associated bone.

Lymphangioma circumscriptum is a rare condition characterized by small groups of vesicles composed of dilated lymphatics in the superficial dermis. It may be present at birth or begin in infancy or childhood.

Diffuse lymphangiomas are poorly circumscribed tumors occurring in the skin, mucous membranes or muscles. They are usually congenital. In the tongue and lips they are responsible for macroglos-sia and macrocheilia, respectively. Diffuse lymphangioma of an extremity is responsible for one form of elephantiasis (elephantiasis lymphangiectatica). It may involve an entire extremity or only a portion, e.g., the fingers or foot; there may be associated hypertrophy of the bone as well as of the soft tissues of the length as well as the girth of the extremity. Histologically the dilated lymphatic channels may be obscured by abundant fibrous tissue. The more localized lesions should be treated by surgical excision, but recurrences are frequent. Treatment of the more diffuse lesions is apt to be unsatisfactory.

Cystic lymphangiomas (cystic hygromas) are most frequently encountered in the neck (hygroma colli) and in the axillae, but may also occur in the inguinal and retroperitoneal regions. In the cervical region they may extend into the mediastinum, and rarely mediastinal hygromas may occur in the absence of a cervical component. Mesenteric cysts are simply cystic lymphangiomas of the mesentery, and many omental cysts are lymphangiomas. Sacral hygromas may simulate lipomas or sacrococcygeal teratomas; they are sometimes connected with the spinal canal. Enlargement of cystic lymphangiomas may occur by enlargement of the individual cysts, by formation of new cysts or by hemorrhage into the cysts.

TUMORS OF THE SOFT TISSUES

Neoplasms arising from muscle, fat and connective tissue comprise a miscellaneous but important group of tumors of early life. They may arise at almost any site and vary from benign neoplasms such as the lipoma to highly malignant sarcomas; the latter may be so undifferentiated as to preclude accurate determination of the cell of origin. They may occur at any age. The most frequent manifestation is a visible or palpable mass. Clinical differentiation of benign and malignant neoplasms is often impossible. Every solid mass should be considered malignant until proved otherwise by histologic examination of the excised mass. **Lymphosarcoma** is the most common malignant neoplasm of the gastrointestinal tract in early life. The tumor usually arises in the small intestine, especially the ileum; it may originate in the colon, appendix or even the stomach. The presenting complaint is usually crampy abdominal pain, often accompanied by vomiting and a palpable mass; the mass may be the neoplasm or an intussusception. Morphologically a segment of the bowel may be diffusely infiltrated by neoplastic cells with resultant thickening of the wall and superficial ulceration of lymph nodes should be removed for diagnostic purposes. Removal of the pancreatic tumor is usually unsuccessful because of its multifocal nature. If the diagnosis of an islet cell tumor can be established histologically either from a primary or a metastatic site in a person with this syndrome, total gastrectomy is probably the treatment of choice, even in children. This may result in disappearance of metastatic lesions.

MALIGNANT TUMORS OF BONE

By far the most frequent primary malignant tumors of bone are the osteosarcoma and Ewing's tumor, the majority of which occur between 10 and 25 years of age; males are affected more frequently than females. Osteosarcoma characteristically involves the metaphyseal end of a long bone, whereas Ewing's tumor involves the shaft, but roentgenographic differentiation of these neoplasms is not always possible. Of greater importance, however, is the fact that many, if not all, of the roentgenographic features of these tumors may be duplicated by non-neoplastic lesions of bone. Accordingly, treatment of lesions suspected of being malignant should not be instituted

until an unequivocal diagnosis is established by histologic study of the tumor. Moreover, the pathologist is limited in his ability to establish a diagnosis on the basis of histologic studies alone. For example, an actively growing callus about a fracture may closely simulate the histologic appearance of an osteosarcoma, yet correlation of the material obtained at biopsy with the roentgenographic findings may clearly indicate the true non-neoplastic nature of the process. **Thus, the pathologist must evaluate all pertinent clinical, roentgenographic and surgical data before he arrives at a diagnosis.**

Osteosarcoma (osteogenic sarcoma) is more common than Ewing's tumor. It usually begins at the lower end of the femur or the upper end of the tibia or humerus, but may arise at other sites. The presenting complaint is commonly that of pain and swelling of the affected part, which the patient may attribute to trauma.

Roentgenographic studies reveal varying degrees of destruction of bone and of new bone formation. Codman's triangle is a radiopacity at the end of the tumor where the periosteum has been elevated. Neither this finding nor the perpendicular striations of new bone in the subperiosteal neoplasm ("sunray appearance") are always present, nor are they pathognomonic of an osteosarcoma. The level of serum alkaline phosphatase may be elevated.

The neoplasm occupies the medullary cavity and penetrates the cortex to the subperiosteal zone; penetration of the periosteum into adjoining soft tissues may also occur. Histologically the appearance is varied, but consists essentially of atypical mesenchymal cells with varying degrees of formation of collagen, typical or atypical osteoid tissue and true bone. Cartilaginous areas and areas of myxomatous tissue may be present. Osteosarcoma commonly metastasizes to the lungs, although other organs may also be involved; osseous metastases are rare. Amputation appears to offer the best possibility of cure, but the case fatality rate is high. In one series an exceptional 5-year survival rate of 19 per cent has been recorded. Recently the use of one or more chemotherapeutic agents has given encouraging results in the control of metastases and probably should be utilized postoperatively in all patients; such therapy can be administered only in a hospital equipped with facilities to control the adverse side effects of the treatment.

Ewing's tumor may involve the same bones as does osteosarcoma; in addition, there is relatively frequent involvement of the flat bones and the ribs. The initial complaints are often similar to those associated with an osteosarcoma; fever and leukocytosis may occur with either tumor, but are more likely to be associated with Ewing's tumor.

Roentgenographically there is a mottled area of rarefaction, often associated with increased density and periosteal formation of new bone. The latter may be deposited in layers, resulting in an "onion-skin" appearance, but this finding is often absent and may appear in association with other osseous lesions. The roentgenographic appearance may closely simulate that of osteomyelitis, osteosarcoma, eosinophilic granuloma of bone or metastatic neuroblastoma.

Gross examination of an affected bone usually reveals more extensive neoplastic involvement than was demonstrable roentgenographically. Histologically the tumor consists of sheets of uniform round or oval nuclei with little or no cytoplasm. The neoplastic cells do not form new bone. Extensive areas of hemorrhage and necrosis are commonly present. The histologic appearance may simulate that of a malignant lymphoma or a metastatic neuroblastoma, and every attempt should be made to exclude the presence of an extraosseous primary lesion.

Ewing's tumor usually involves a single bone when first recognized, but ultimately many bones may be affected. Metastases to the lungs are also common. Treatment currently consists in supervoltage irradiation and chemotherapy rather than amputation. The 5-year survival rate should be in the vicinity of 25 per cent or possibly more; occasionally apparent cures have been effected after the development of metastases.

TUMORS OF THE KIDNEY

Wilms' tumor (nephroblastoma) is one of the most common abdominal neoplasms of early life; approximately two thirds appear before the age of 4 years. Although authentic Wilms' tumors do occur in the neonatal period, almost all the tumors reported as such at this early age have been leio-myomatous hamartomas of the kidney; either of these occurring at birth may be

accompanied by polyhydramnios. Bilateral renal involvement is uncommon; when it occurs it is usually detectable at the time of the initial diagnosis; otherwise, the second tumor usually appears within a year or so. Wilms' tumor occurs with increased frequency in children with aniridia, hemihypertrophy and Beckwith-Wiedemann syndrome and probably in those with fused kidneys. The incidence of Wilms' tumor with bilateral aniridia has been reported as high as 1:73 as contrasted with the usual rate of 1:50,000 to 1:100,000 and conversely, Wilms' tumors have occurred in as many as 7 of 28 children under 4 years of age hospitalized with aniridia. The aniridia accompanying Wilms' tumor is almost always of the sporadic type and is apt to be associated with other congenital defects, e.g., cataracts, mental retardation and genitourinary anomalies, including cryptorchidism.

The presenting complaint is usually that of an abdominal mass. Abdominal pain and fever may be the first manifestation. Hematuria is relatively infrequent and, contrary to earlier opinions, is probably not a poor prognostic sign. Physical examination reveals a firm, nontender mass in the renal area which may extend down into the iliac fossa but usually does not cross the midline. Hypertension, the pathogenesis of which is not always clear, may be present.

Roentgen examination reveals a soft tissue density which is apt to displace the intestine toward the opposite side. Calcification is infrequent; when present it is apt to be dense and curvilinear, in contrast to the stippled appearance common in a neuroblastoma. Pyelography, preferably including an inferior vena cavagram, usually reveals distortion of the renal pelvis; displacement or extension of the tumor into the inferior vena cava is sometimes apparent. In some instances the kidney on the affected side cannot be visualized by intravenous pyelography. The pyelographic findings are not pathognomonic of a Wilms' tumor but instead indicate the presence of an intrarenal mass. Probably the most important reason for obtaining pyelo-grams is the demonstration of the presence and apparent normality of the opposite kidney.

Macroscopically the tumor usually presents as a bulky circumscribed mass replacing much of the affected kidney and covered externally by the thin renal capsule. With continued growth, however, there may be invasion of the renal pelvis and/or veins and extension beyond the capsule into the perirenal fat, adrenal, diaphragm or colon. On section the tumor bulges beyond the surface of the adjoining kidney. It is yellowish gray and soft, friable or semiliquid as a result of multiple areas of necrosis; it often contains myxomatous areas and foci of hemorrhage. Areas of necrosis may be responsible for cysts containing clear or hemorrhagic fluid. The renal pelvis is usually elongated and distorted, and occasionally masses of neoplastic tissue extend into its lumen. Careful search should be made for islands of neoplastic tissue in the remainder of the affected kidney since there is some evidence to indicate that such multicentric or metastatic lesions affect the prognosis adversely. Metastases occur principally in lymph nodes, lung and liver.

The histologic pattern is variable. There are often broad sheets or cords of undifferentiated mesenchymal cells within which are scattered epithelial-lined tubules; rarely the latter, if incompletely differentiated, may resemble the pseudoro-settes of a neuroblastoma. Abortive glomeruli are sometimes present. Bands of loose, more differentiated mesenchymal tissue tend to divide the more cellular areas into coarse lobules, and within these bands smooth and skeletal muscle or, less frequently, bone or cartilage may be present. There is some evidence to suggest that those patients in whom the neoplasm is predominantly epithelial have a better prognosis.

The basic treatment is prompt (but not emergency) radical nephrectomy performed under optimal conditions, usually within 24 to 48 hours after discovery of the mass. Undue palpation of the tumor should be avoided. Preoperatively, roentgen films of the chest should be obtained and the presence and apparent normality of the opposite kidney established by pyelography. Compatible blood should be available if needed for transfusion during operation. At the time of surgery, which should be by the transabdominal approach, the contralateral kidney should be carefully inspected and palpated, the abdominal viscera investigated for evidence of neoplasm and extent of the tumor marked by radiopaque clips.

An attempt to determine optimum therapy is currently being conducted by the National Wilms' Tumor Study. The patients are grouped as follows:

Group I: The tumor is limited to the kidney and is completely resected.

Group II: The tumor extends beyond the kidney but is completely resected; e.g., there has been penetration beyond the capsule, involvement of peri-aortic lymph nodes or infiltration of renal vessels outside the kidney, but there is no apparent tumor beyond the margins of resection.

Group III: There is residual nonhematogenous tumor confined to the abdomen. This includes those patients in whom biopsies have been taken of the tumor, in whom the tumor has ruptured before or during surgery, in whom there are local peritoneal implants, nodal involvement beyond the periaortic chain, or in whom the tumor cannot be completely resected.

Group IV: There are hematogenous metastases, e.g., to the lungs, liver, bone or brain.

Group V: There is bilateral renal involvement either apparent at operation or appearing subsequently.

The treatment of all patients entered into the study is randomized. All patients in Group I are treated by excision and the administration of actinomycin D and vincristine; irradiation is no longer routinely used. All patients in Groups II and III are treated by excision, irradiation, actinomycin D and vincristine. Randomization is currently employed in an attempt to determine the optimum duration of chromotherapy which might be utilized in order to effect cure and to avoid any sequelae.

The results of this collaborative study are not yet available, but the improvement in prognosis following excision, irradiation and chemotherapy has been striking. In one series apparent cures were obtained in 47 of 53 patients who had no demonstrable metastases on admission and in 18 of 31 of those with metastases. With rare exceptions, if a child with a Wilms' tumor is alive and well with no evidence of recurrence or metastatic disease two years after removal of the tumor, a cure has been effected. Such patients should be followed indefinitely, however, for the possible development of subsequent neoplasms secondary to the effects of irradiation.

TUMORS OF THE ADRENAL

Neuroblastoma is one of the most common malignant neoplasms in infants and children. Although more than half of them arise from the adrenal or from the retroperitoneal sympathetic chain, the neoplasm may originate at any site along the sympathetic chain, e.g., in the posterior mediastinum, pelvis or cervical sympathetic ganglia; the tumor may also arise from other derivatives of neural crest origin, such as the dorsal root ganglia. Neuroblastoma is primarily a disease of early life; one fourth of the affected persons have their initial manifestations during the first year of life and three fourths before the age of 5 years. It is the most common malignant neoplasm to be identified at birth, and metastases may even be present at this time.

The presenting manifestation of a neuroblastoma arising in the adrenal or in the neighboring sympathetic ganglia is usually an abdominal mass. It often crosses the midline, in contrast to Wilms' tumor. Roentgen examination reveals a soft tissue mass which displaces the kidney on the affected side downward and laterally; focal areas of calcification are often present. Intravenous pyelography characteristically reveals displacement rather than distortion of the renal pelvis; occasionally, as with Wilms' tumor, the pelvis is not visible on the affected side.

Intrathoracic neuroblastomas are almost always located in the posterior portion of the mediastinum at any level and may be responsible for cough, dyspnea and pain in the chest. A mass may be responsible for separation of the posterior portions of the ribs and some narrowing and erosion of them. In a number of instances, owing to extradural extensions of the mass, there are manifestations referable to compression of the spinal cord. **Pelvic neuroblastomas** usually produce a demonstrable mass which in some instances simulates a sacrococcygeal teratoma. They may be responsible for urinary or rectal obstruction. **Cervical neuroblastomas** usually do not reach a large size before being recognized. They are apt to present as a hard, lobulated mass involving the posterior triangle of the neck or extending both anterior and posterior to the

sternocleidomastoid muscle. In some instances fine stippled areas of calcification within the mass are demonstrable roentgenographically.

The majority of patients with neuroblastoma have metastases when the tumor is first recognized. In some instances the presenting complaint is referable to metastases rather than to the primary tumor, e.g., massive hepatomegaly, especially in young infants, or cervical or axillary lymphadenopathy. Persistent pain and fever may occur with osseous metastases even in the absence of roentgenographic changes and may simulate rheumatic fever or rheumatoid arthritis. Involvement of retrobulbar soft tissues, probably secondary to osseous metastases, may be responsible for proptosis and/or ecchymosis of the upper eyelids. One or more bluish subcutaneous nodules may precede other complaints, especially during the neonatal period. Infrequently, only the metastatic disease is apparent during life, no primary site being identified.

The majority of patients with neuroblastoma have elevated levels of catecholamines or of one or more of their derivatives in the urine. There is some evidence to suggest that this may not occur with tumors which arise in the dorsal root ganglia. Relatively few patients with neuroblastoma, however, initially present with signs of functional endocrine activity such as flushing, perspiration, tachycardia and headache. Intractable diarrhea is an uncommon manifestation. Measurements of the urinary excretion of catecholamines and of their metabolites, e.g., 3-methoxy-4 hydroxymandelic acid (VMA) and homovanillic acid (HVA) may be of diagnostic significance and may also be helpful in the demonstration of residual, recurrent or metastatic disease. Cystathioninuria may also be a diagnostic aid but is less reliable.

Uncommonly acute cerebellar encephalopathy may precede, follow or occur concomitantly with the discovery of a neuroblastoma. There are ataxia, weakness of the extremities and oculogyric crises unaccompanied by pleocytosis in the cerebrospinal fluid and with little or no fever. The cerebellar signs may disappear following removal of the tumor, but mental retardation may persist. Although the mechanism responsible for this association is not clear, patients with acute cerebellar encephalopathy should probably be investigated for the possible presence of an inapparent neuroblastoma.

Metastases of neuroblastoma occur by way of the lymphatic and blood streams; regional and distant lymph nodes, the skeletal system and the liver are the most frequent sites of metastatic spread. Pulmonary metastases occur in only about 10 per cent of patients. Osseous metastases are often bilateral; a unilateral lesion may lead to an erroneous diagnosis of a primary neoplasm of the bone. The roentgenographic changes in the skeleton are characterized by areas of destruction and proliferation of new bone, which may closely simulate the appearance of Ewing's tumor, of eosinophilic granuloma of bone or of skeletal involvement in leukemia. There may be extensive mottling of the cranial bones and separation of the sutures, owing to increased intracranial pressure from metastatic invasion of the dura mater. Neoplastic cells are frequently demonstrable in smears of the bone marrow, even in the absence of roentgenographic changes in the bones themselves.

The neuroblastoma is initially an encapsulated neoplasm, but it soon infiltrates adjoining tissues and, if arising in the adrenal or neighboring sympathetic ganglia, may surround the aorta, inferior vena cava, ureter or renal pedicle and render complete surgical removal impossible. Areas of hemorrhage and necrosis are commonly present, as are minute flecks of calcium. Histologically there may be varying degrees of differentiation toward mature ganglion cells or, less frequently, toward chromaffin cells. The least differentiated neoplasms may be misinterpreted as lymphosarcomas, but additional sections of the same tumor will usually reveal better differentiated areas of neoplastic cells embedded in a haphazard manner within a delicate fibrillary tissue or arranged as pseudorosettes. Less frequently, immature or mature ganglion cells or even chromaffin cells may be present.

The prognosis for the child with a neuroblastoma is dependent upon a number of factors, e.g., extension of the neoplasm across the midline and the extent of maturation of the tumor as determined histologically. The poorer prognosis related to abdominal tumors as contrasted with cervical or mediastinal ones is probably largely dependent upon the more advanced stage of the

disease at the time of diagnosis. Age appears to be the single most important factor with respect to prognosis. The over-all survival rate is in the range of 30 per cent and increases to about 70 per cent in those under 2 years of age. Infants less than 1 year of age, even with metastases to the liver, skin and/or bone marrow but without roentgenographic changes in the bones, have good chances of survival. Cures in the presence of demonstrable osseous metastases have been observed infrequently, even in children over 2 years of age.

Complete surgical removal of the primary tumor is the treatment of choice, but even incomplete removal may be followed by a cure; irradiation is probably indicated if all the tumor is not removed. Chemotherapy, especially with vincristine sulfate and cyclophosphamide (Cytoxan) may cause striking regression of the tumor and relief of symptoms in children with widespread disease, but its role in the treatment of those with more localized disease has yet to be established. Certainly the known adverse effects of irradiation and the possibly still unknown effects of chemotherapy must be considered in evaluating the optimum therapy for any patient with this unpredictable neoplasm, in which even spontaneous cures may take place.

Sacrococcygeal teratomas arise from the region of the coccyx or lowermost part of the sacrum. They are probably derived from the primordial, to-tipotential cells of the primitive knot (Hensen's node) which, during embryonic life, finally comes to rest in the region of the coccyx. These tumors are three or four times more frequent in girls than in boys, and there is a significant increase in the incidence of twinning in families of persons with sacrococcygeal teratomas. At least three fourths of the tumors are apparent at birth, usually presenting as a mass at the tip of the coccyx extending externally in the midline or into one or both buttocks. Large tumors, which may exceed the size of the infant's head, displace the coccyx posteriorly and the anus anteriorly. Occasionally the mass is responsible for urinary or intestinal obstruction, but, in contrast to large myelomeningoceles, they are not responsible for neurologic defects in the extremities.

Rectal examination usually discloses a readily; palpable mass posterior to the rectum, which is sometimes encircled by it. Roentgenographic examination usually reveals a soft tissue density in the pelvis, sometimes with displacement of the coccyx posteriorly. Areas of calcification or actual bone are demonstrable in about half of the tumors, In contrast to sacral chordomas, roentgenographic evidence of destruction of the sacrum is rare; when present, it is indicative of a malignant neoplasm. Occasionally spina bifida or lumbosacral anomalies are also present. The differential diagnosis includes meningocele or meningomyelocele; pressure on such a sac will cause the fontanel to bulge, or, if this is closed, crying or straining should increase tension within the mass. Neurogenic tumors, e.g., neuroblastoma and ganglioneuroma, may be clinically indistin-guishable from a sacrococcygeal teratoma. Chordomas are rare in children; they are responsible for destruction of the sacrum and only rarely extend into the buttock. Papillary ependymoma (below) may present as a mass in the sacrococcygeal region. Cystic lymphangiomas and hemangiomas may simulate sacrococcygeal teratoma, as may duplication of the hind gut. Occasionally a sacro-coccygeal teratoma presents as a red, inflamed mass or as a draining sinus and thus simulates an infected pilonidal sinus.

The neoplasms are connected to the lowermost part of the sacrum or to the coccyx; the coccyx should always be removed with them. Rarely the tumor extends into the vertebral canal. The tumors are usually well circumscribed solid masses containing multiple cystic structures. Histologically they contain a vast array of tissues; fat, neural elements, smooth and skeletal muscle, bone, cartilage and intestinal and bronchial elements are the most frequent. Teratomatous elements such as pancreatic islets or adrenocortical tissue are sometimes present and may, rarely, produce functional manifestations. Most sacrococcygeal teratomas are benign, and cures are sometimes obtained even after one or more recurrences. Most of the tumors discovered before 2 months of age consist only of mature or less often of immature fetal elements and are associated with an excellent prognosis, whereas those appearing later usually contain embryonal carcinomatous areas and are highly malignant. Tumors present at birth but not excised until after 4 months of age are more apt to be malignant

than those removed earlier. Symptoms of bowel or bladder dysfunction increase the probability that the neoplasm is malignant. Tumors detected after 5 years of age, as in adults, may be benign or malignant. Recurrences or metastases, if they occur, usually do so within two years after operation.

TUMORS OF THE MEDIASTINUM

Mediastinal masses in infants and children are relatively common. If malignant lymphomas are excluded, most of which are accompanied by manifestations in addition to those referable to the mediastinum, approximately three fourths of the mediastinal masses are neurogenic or teratomatous neoplasms or non-neoplastic cysts, e.g., duplications of the esophagus and neurenteric or bronchogenic cysts. Approximately 25 per cent of mediastinal masses in infants and children are malignant as compared with 15 per cent of those in adults. Tumors arising in the anterior mediastinum are predominantly teratomas, whereas most of those originating in the posterior mediastinum are neurogenic neoplasms; masses confined to the mid-mediastinum are usually lymphomas or nonneoplastic cysts.

Approximately two thirds of the infants and children with a mediastinal mass are symptomatic; in the others the lesion is a chance finding on a roentgenogram of the chest. Cough, dyspnea, stridor and pain are the most frequent manifestations and are especially apt to occur with teratomas, malignant neoplasms of any type and with nonneoplastic cysts; vascular tumors and benign neurogenic ones are often unassociated with respiratory symptoms.

Mediastinal teratomas are located in the anterior mediastinum, usually in its superior aspect. Many of them apparently arise in the thymus. Rarely they arise within the pericardial sac and simulate a congenital cardiac lesion. Many are benign cystic neoplasms, commonly referred to as **dermoid cysts,** which are cystic variants of the more solid teratomas. Symptoms may not be apparent until adult life. Dyspnea, cyanosis and cough may be manifestations, and expectoration of hair and sebaceous material may occur if the tumor perforates into a bronchus. Infection of the cystic mass may produce symptoms simulating a pneumonic process. Rarely the neoplasm extends into the suprasternal or supraclavicular area. Compression of the superior vena cava causes dilation of the veins of the head, neck and upper part of the thorax. Roentgenograph^ examination reveals a circumscribed mass extending from the anterior mediastinum into one hemithorax; when teeth or skeletal elements are demonstrable roentgenographically, the nature of the mass is established.

The teratomas may be composed of one or more cysts; less frequently they are predominantly solid tumors. The cysts contain sebaceous material, hair or mucoid material. Histologically almost any type of tissue may be present, especially in the solid neoplasms. Malignant teratomas, nearly all of which occur in males, are usually solid or finely cystic tumors containing actively proliferating, poorly differentiated tissue in addition to more mature elements; metastatic lesions may resemble the primary tumor or consist only of embryonal carcinoma. Mediastinal teratomas should be surgically removed.

Thymomas are rare in children. In adults they sometimes accompany or precede the development of myasthenia gravis. This association is extremely rare in children, as is the association of thymoma with aregenerative anemia. Thymomas may be asymptomatic and discovered only roentgenographically, or they may be responsible for vague retrosternal pain, cough, dyspnea or signs of compression of the superior vena cava. They are usually encapsulated and composed of an admixture of lymphoid and epithelial cells; typical Hassall's corpuscles are rare. True thymomas are usually benign; occasionally they infiltrate and implant on the pleura.

Lymphosarcoma is the most common malignant neoplasm of the gastrointestinal tract in early life. The tumor usually arises in the small intestine, especially the ileum; it may originate in the colon, appendix or even the stomach. The presenting complaint is usually crampy abdominal pain, often accompanied by vomiting and a palpable mass; the mass may be the neoplasm or an intussusception. Morphologically a segment of the bowel may be diffusely infiltrated by neoplastic cells with resultant thickening of the wall and superficial ulceration of lymph nodes should be removed for diagnostic purposes. Removal of the pancreatic tumor is usually

unsuccessful because of its multifocal nature. If the diagnosis of an islet cell tumor can be established histologically either from a primary or a metastatic site in a person with this syndrome, total gastrectomy is probably the treatment of choice, even in children. This may result in disappearance of metastatic lesions.

5. Additional materials for the self-control

A. Clinical cases

Case 1. The mother complains about the enlarging of the abdomen of her eighteen-month old child, which she noticed two weeks ago, the child is inert, and has poor appetite. At the examination: the skin of the child is pale, the abdomen is enlarged, asymmetric. During the palpation of the abdomen the tumour in the projection of the left kidney was detected, it is dense, not displaced. The blood test showed anaemia, in the urinalisis –3-5 red cells. What diagnosis have you made? What tactics have you developed?

Case 2. In the chamber of the maternity hospital you examined a new-born child and detected a tumour in the area of coccyx by the size of the head of a new-born one. The skin above the tumour is of the normal color. During the palpation the tumour is heterogeneous, not mobile. There is no dysfunction of the pelvic organs. What diagnosis have you made? What tactics have you developed?

Case 3. A 6 month-old boy was hospitalized into the surgical department with the complaints of the mother about a tumular neoplasm in the area of the back. The child has had the tumour since his birth and it increases slowly. At the examination: the tumour of the red color 2x3 cm is detected in the right lumbar area, while being ptressed the tumour turns pale and diminishes in size. What diagnosis have you made? What tactics have you developed?

Case 4. At the examination of an 11 year-old child with the kidney tumor under suspicion it was detected on the excretory urography that the left kidney is displaced and deformed, and the ureter is displaced in the medial direction. At the irrigography it was detected that the tumular neoplasm displaces the sigmoid colon forward and medially. At the USD a dense formation of heterogeneous structure is detected in the left half of the abdominal cavity from the left hypochondrium to the entrance into the small pelvis, which is located in the retroabdominal space. What diagnosis have you made? What tactics have you developed?

Case 5. The survey sciagram of the chest of a 14 year-old child shows a semispherical shade 50×45 mm to the right that adjoins with its wide basis to the shade of the mediastinum, as if blending with it by its internal contour, the external contour of the shade is clear enough. What diagnosis have you made? What tactics have you developed?

Case 6. A 3 year-old girl which has been ill for a month complains about a considerable loss of weight, intoxication, anaemia. At the examionation on the excretory urography conducted on the background of the pneumoperitoneun, it was detected that the whole right half of the abdominal cavity is filled with a tumour, the inestine is displaced to the left. The function of the left kidney is not changed, the function of the right kidney is absent. The sciagram of the lungs doesn't show any pathological changes. What diagnosis have you made? What tactics have you developed?

B. Tests

6. LITERATURE FOR STUDENTS

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