Clinical Observation: Malignant Schwannoma of the Retroperitoneal Space.

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ABSTRACT

The article presents a clinical case of malignant retroperitoneal schwannoma, which was a diagnostic finding. The clinical picture of the disease was dominated by pain in the lower abdomen and lower back, frequent urination. After a diagnostic examination and preliminary preparation, the patient was operated on in a hospital at the oncogynecology department of the Tashkent City branch of the Republican Specialized Scientific and Practical Medical Center of Oncology and Radiology with a preliminary diagnosis of an ovarian tumor. Interrooperatively, a tumor of the retroperitoneal space with lesions of the obturator nerve was revealed. Subsequently, immunohistochemical (IHC) analysis clarified the organ belonging and the histological type of the tumor, which turned out to be a malignant schwannoma of the retroperitoneal space. These data may help in the management of patients with this pathology and improve the diagnosis of patients with pelvic tumors.

Keywords: malignant schwannoma, retroperitoneal tumor, differential diagnosis of ovarian tumor, IHC.

Introduction. Schwannomas, also known as neurilemmomas, are tumors composed of well-differentiated Schwann cells derived from the glial cells of peripheral nerve sheaths [1]. Most

schwannomas are benign, and malignant ones, which are usually associated with von Recklinghausen's disease, are rare [2,3]. Schwannomas usually occur in the head and neck, retroperitoneum, and extremities. The pelvic form is very rare, with a reported incidence of 1-3% of all schwannomas [4,11,12]. Since there are no specific clinical or radiological signs for pelvic schwannomas, and they resemble a number of diseases of the pelvic organs, an erroneous diagnosis can be easily made [5,6]. Surgical excision is both diagnostic and therapeutic in the treatment of pelvic schwannomas. In 50% of cases, schwannomas are combined with neurofibromatosis. The frequency of local recurrence is about 40%, and in 30-60% there are distant metastases to other organs such as the lungs, bones and pleura. Clinical and morphological features of retroperitoneal tumors are due to the anatomical structure of the retroperitoneal space, the significant prevalence and diversity of tissue structures in this area. The latter determines the variety of histological forms of tumors that have different morphological and functional properties that determine their clinical course [3]. These tumors, especially poorly differentiated ones, grow relatively rapidly, recur early and give distant metastases.

Due to the rarity of this oncopathology, we present a clinical case that may be of particular interest to surgeons and pathologists.

Patient Z., 66 years old, was admitted to the oncogynecology department of the Tashkent city branch of the Republican Specialized Scientific and Practical Medical Center of Oncology and Radiology (TCB RSSPMCOandR) 11/14/22 with complaints of recurring pain in the lower abdomen and lower back, frequent urination. From the anamnesis he has been ill for 4 months. In July 2022, the patient began to have the above complaints, underwent ultrasound of the pelvic organs. Ultrasound revealed ovarian masses. Then, she turned to the clinic at the place of residence, examined by a gynecologist, and a consultation with an oncologist was recommended. Subsequently, the patient was referred to TCB RSSPMCOandR, where she was examined by an oncogynecologist, additionally examined. Tumor markers from 05.11.22: CA125 - 12.0 U / ml, HE4 68.7rM, ROMA index 13.3%. MRI (abdominal cavity, retroperitoneal space, small pelvis) dated 11/09/2022: Conclusion: MRI signs of left-sided urethrohydronephrosis, uterine fibroids. Volumetric formation of the ovary.

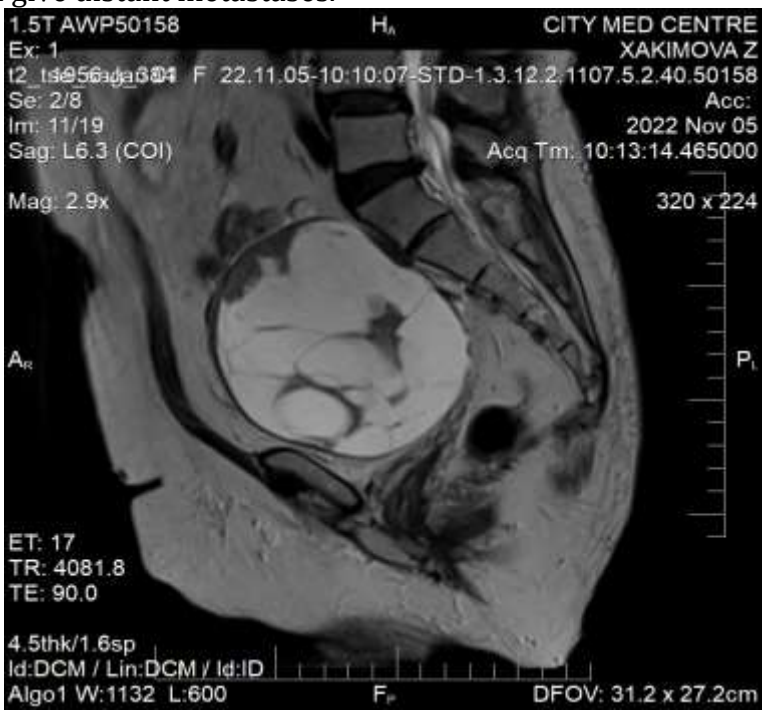

Figure 1. MRI examination of the pelvis. MRI - signs of uterine fibroids. Volumetric formation of the ovary (sagittal section).

Subsequently, the patient was diagnosed with Suspicion of a tumor of the left ovary. Complications: a symptom of pain. Urethrohydronephrosis on the left. The patient is hospitalized in the department of oncogynecology for further examination and decision on further treatment tactics.

On admission, the patient's general condition was satisfactory. Skin and visible mucous membranes of normal color. There is no hyperthermia. Breathing is vesicular, it is carried out in all departments, there are no wheezing. Respiratory rate is 16 per minute. Rhythmic heart sounds, BP 130/80 mm Hg. Art. Pulse 78 per minute. The tongue is clean and moist. The abdomen is not swollen, soft on palpation, painless in all departments, participates in the act of breathing. Peristalsis is heard. There are no peritoneal symptoms. The effleurage symptom is negative on both sides. The feces is adequate. Urination is frequent, in small portions, light urine.

According to the laboratory examination, an increase in ESR (erythrocyte sedimentation rate) to 19 mm/h, a decrease in hemoglobin to 96 g/l, a slight increase in erythrocyte-, leukocyte- and proteinuria, a slight increase in blood creatinine to 114 μmol/l were revealed. Ultrasound: in the abdominal cavity, retroperitoneal space, as well as in the small pelvis, free fluid and delimited fluid accumulations were not detected. The liver, biliary tract and pancreas are not changed, the structure of the spleen is homogeneous, the right kidney is not structurally changed, the left kidney is 114x56 mm in size, the parenchyma is uniformly thinned to 5 mm, increased echogenicity, severe calicopyeloureterectasia (pelvis 56 mm, calyx up to 53 mm); In the small pelvis, more to the left in the projection of the left ovary, an irregularly shaped formation is determined, measuring 181.2x99.8x13.7 mm. The formation is heterogeneous in structure, mostly solid (heterogeneously low echogenicity) with the presence of cystic cavities of various shapes and sizes (the contents of some cavities are diffusely heterogeneous). Uterus in RFV, dimensions 65x74x56 mm, myometrium of a homogeneous structure. Endometrium - 3 mm. The right ovary is 30x26 mm in size and has a homogeneous structure. Conclusions: Tumor of the left ovary. Ureterohydronephrosis on the left I degree.

X-ray of the chest organs: fresh focal and infiltrative shadows were not revealed. EchoCG: General and regional contractility of the left ventricle is not broken. The valves and septa of the heart are intact. The cavities of the left ventricle are not dilated. The pericardium is not dilated. Tachyarrhythmia. Diastolic dysfunction of the left ventricle type 1. Doppler of the lower extremities. Conclusion: The main arteries of the vessels of the lower extremities are passable. Hemodynamically significant stenosis was not detected. Cytology no. 105886/22: Puncture of the posterior fornix: Cytology with suspected cell atypization. Additional studies are needed to differentiate and definitively diagnose neoplasms.

Status Genitalis: external genitalia developed correctly, female type of hair. In the mirrors: the cervix is retracted to the right, the discharge is light. Per Vaginum: The body of the uterus and appendages are in a single conglomerate, palpable formations are 15 cm in diameter, with limited mobility. Parameters are free.

According to the results of the examination, the patient was diagnosed with a preoperative diagnosis: Primary: Tumor of the left ovary. Complications: Ureterohydronephrosis on the left, 1st degree. Pain symptom. Concomitant: chronic bronchitis. After ulcerative scar of the duodenum. Catarrhal gastritis.

On 11/23/2022, the operation was performed: LAPAROTOMY, REVISION, EXTIRPATION OF THE UTERUS WITH ADDITIONS, REMOVAL OF THE RETROPERITONEAL TUMOR. The course of the operation: Under epidural anesthesia, a lower median laparotomy was performed in 18 cm long, the thickness of the subcutaneous fat was up to 5 cm. During the revision: there is a huge tumor emanating from the obturator fossa on the left, filling the iliac space with dimensions of 15x20x25 cm, cystic-solid structure, intimately adjacent to the internal and external iliac vein, and the artery, ureter, and the back wall of the bladder. The body of the uterus is enlarged up to 7 weeks of pregnancy, dense - elastic consistency, deviated to the right due to the tumor (Fig. 2).

The cervix is 2.0x4.0 cm in size, hypotrophic. Pipes on both sides are unchanged. The ovaries are cystically altered on both sides, 4x3.5 cm in size. The liver and other organs of the abdominal cavity - without visible pathology. On the round on both sides and funnel-pelvic ligaments on the left, clamps are applied, crossed, stitched and alloyed. The vesicouterine and recto-uterine folds are acutely deflated. Clamps were placed on the cervical vessels on both sides, crossed, sutured and ligated. Produced extirpation of the uterus with appendages. The stump of the vagina is sutured tightly. Hemostasis

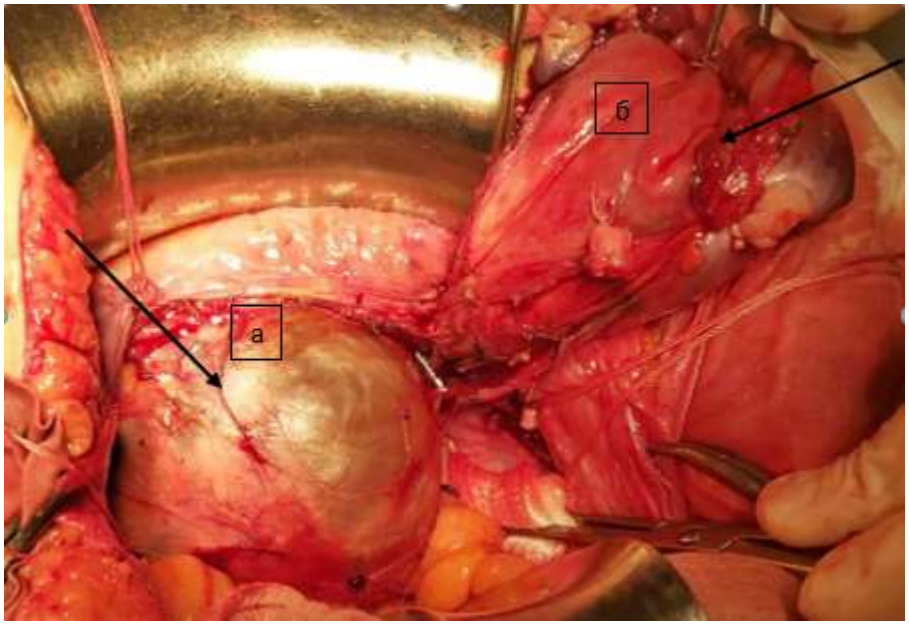

Figure 2. Stages of the operation: a - cystic part of the tumor, b - uterus with appendages.

Started mobilization of the retroperitoneal tumor on the left, with technical difficulties made the separation of large vessels from the tumor, especially the external iliac vein and artery. The tumor grows into the mesentery of the sigmoid colon, the adhesions are dissected in a sharp way, atraumatic gray-serous sutures are applied to the deserized parts of the intestine for 8 cm. Upon further revision, it was found that the tumor was growing into the internal iliac vein and artery (Fig. 3), taking into account the above, the internal iliac artery and vein were crossed, sutured and ligated. The distal part of the ureter on the left was mobilized, and it was revealed that there is an expansion of the ureter. Part of the tumor was sent for express histological analysis. The answer of the urgent histological analysis No. 11589: alveolar rhabdomyosarcoma.

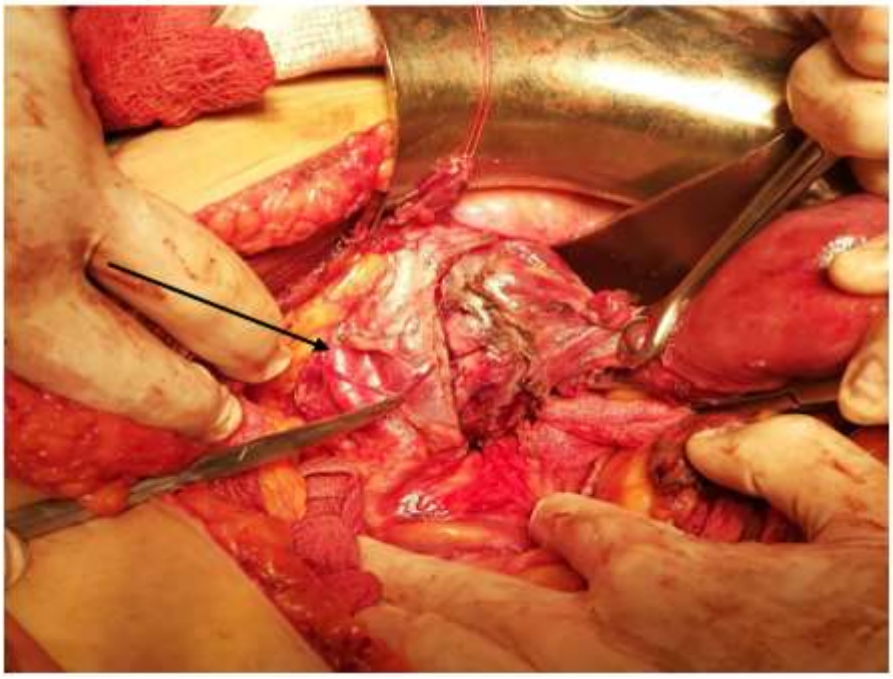

Figure 3. Stages of the operation: detection of tumor invasion into the internal iliac vein and artery.

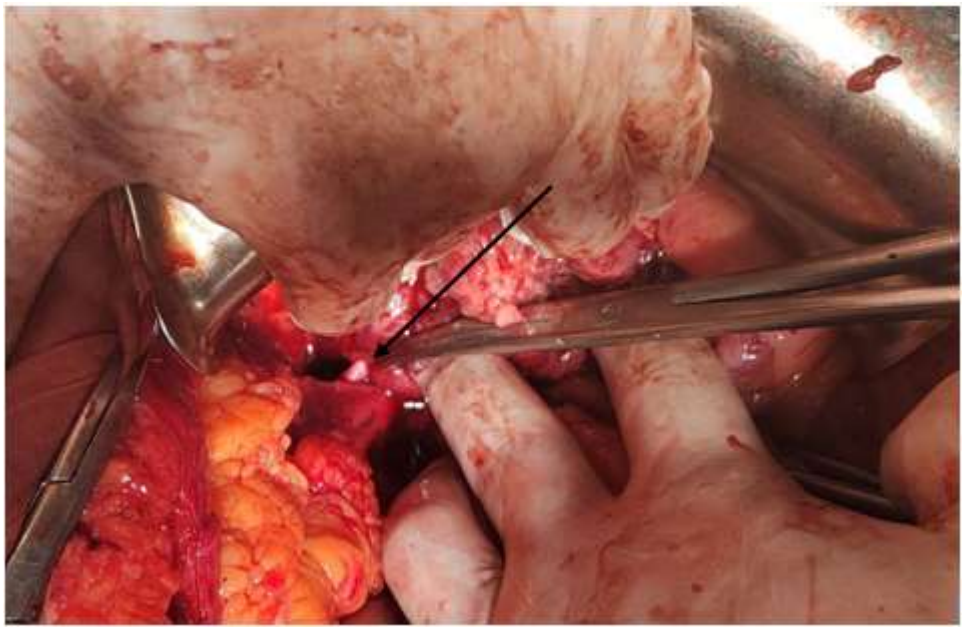

Figure 4. Stages of the operation: the obturator nerve runs inside the tumor.

Upon further revision, it was found that the obturator nerve passes inside the tumor (Fig. 4). Taking into account the germination of the tumor in the obturator nerve and the defeat of this area inside the tumor, it was decided interooperatively to resect the part of the n.obturatorius with the tumor based on the principle of radicalism.

With technical difficulties, the retroperitoneal tumor was removed on the left. The operating bed was washed with aseptic solutions, sanitized (2 liters). Peritonization. Hemostasis. Produced drainage of the pelvis, the drainage tubes are brought out through the wound on the right and left sides. Napkin count. The anterior abdominal wall is sutured in layers, tightly. Urine through the catheter 200 ml, light. Blood loss 450.0 ml.

The course of the postoperative period is smooth. The sutures were removed on the 10th

day. The urethral catheter was removed on the 10th day after the operation, spontaneous urination was restored. On the 12th day after the operation, the patient was discharged in a satisfactory condition.

Histological examination. The surgical material on the cut is a malignant formation closely adjacent to the nerve bundles. Tumor cells are wrapped in sheets. Some cells with a high nuclear cytoplasmic index, with minimal cytoplasm and round nuclei (Fig. 5). A focus of necrosis is noted. Mitotic activity - 1-2/1 field of view. The tumor grows into the surrounding fibrous tissue and one reactive node.

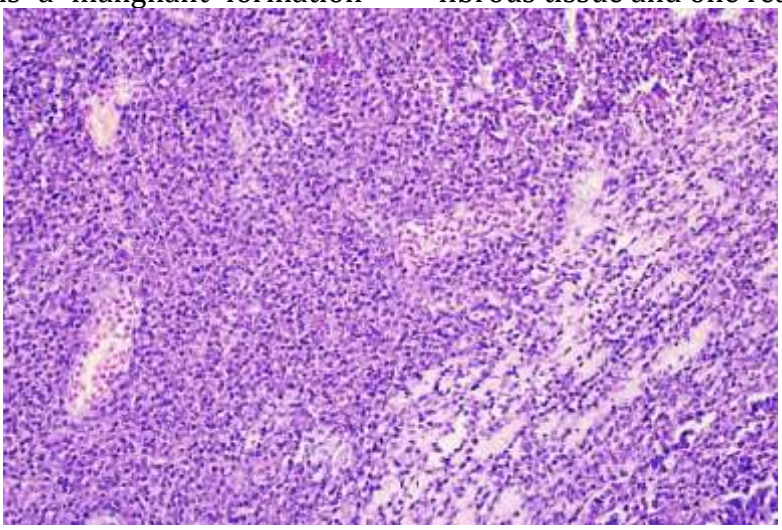

Figure 5. Microscopic picture of the intraoperative material.

In the myxoid stroma with foci of hyalinosis, hypercellular areas are visible, consisting of malignant cells from a round to fusiform structure. Cells with moderate polymorphism, sparse cytoplasm, nuclei with visible vesicular nucleoli.

In a planned histological examination, the morphological picture of a malignant neoplasm had similar features with such tumors as rhabdomyosarcoma, leiomyosarcoma, extraintestinal GIST, synovial sarcoma, myeloid sarcoma, and malignant schwannoma. Neoplasia resembles a biphasic tumor, consisting of alternating foci of various structures. There are layers of closely spaced spindle-shaped structures, as well as small round, blue cells with signs of immature differentiation (Fig. 6)

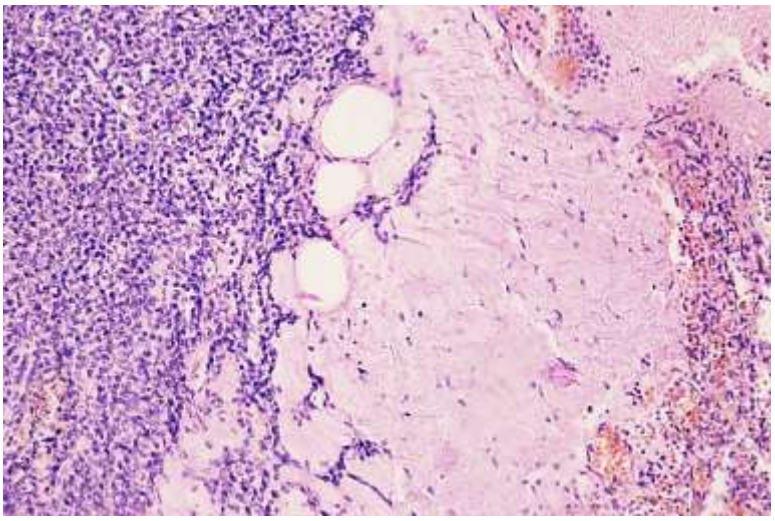

Figure 6. Morphological picture of a poorly differentiated neoplasm. (Staining with hematoxylin-eosin x100)

An immunohistochemical study was performed to clarify the diagnosis. On immunohistochemical profiling of the tumor, Myogenin, MyoD1 and Desmin were all negative and the initial diagnosis of rhabdomyosarcoma was withdrawn. After that, a panel of IHC markers was created, including PanCK, CD117, DOG1, CD34, SMA, BCL2, MPO, TdT, CD99 and S100. To our surprise, PanCK, CD117, TdT, CD99 stained small areas in the tumor tissue. Since CD34 and DOG1 were negative, GIST was not confirmed. Despite focal TdT and CD99 positivity, MPO was negative, and the diagnosis of myeloid sarcoma was also ruled out. Biphasic synovial sarcoma and malignant schwannoma remained for the differential diagnosis. Given the diffuse and strong expression of S-100 and BCL2 negativity, synovial sarcoma was also excluded. Additional markers were placed to rule out melanoma. HMB45, MelanA and SOX10 were negative. Immunoprofiling of the resected tumor corresponded to poorly differentiated malignant schwannoma (Fig. 7).

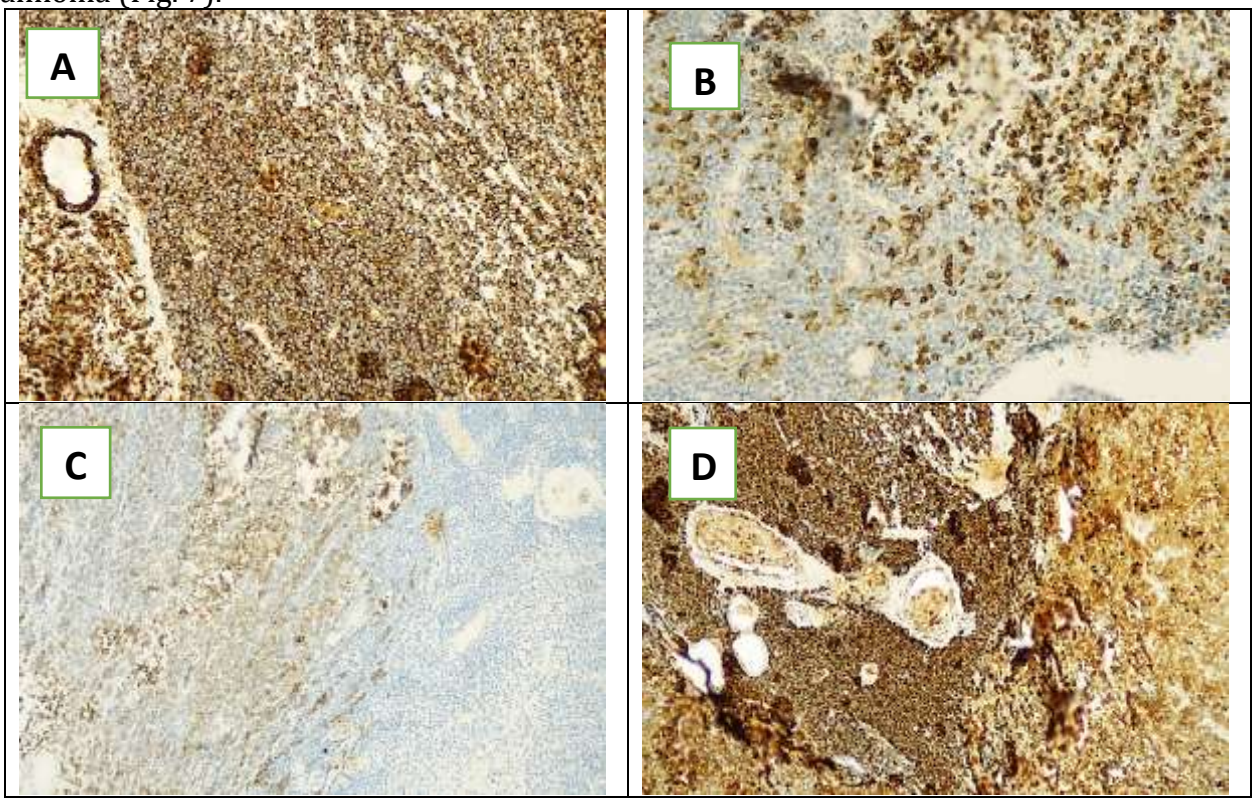

Figure 7. IHC pattern of poorly differentiated malignant schwannoma. A. Diffuse positive expression of Vimentin (IHC staining, x100). B. Focal positive expression for PanCK (IHC staining, x100). C. Focal positive expression for CD117 (IHC staining, x100). D. Diffuse positive reaction to S100 (IHC staining, x100).

Discussions. Malignant schwannoma, or the so-called malignant neurolemmoma, develops from the Schwann sheath of the nerve and accounts for less than 5% of all malignant soft tissue formations. By localization, malignant neurolemma most often affects the nerve plexuses of the upper half of the trunk, neck, upper limbs, small pelvis, less often localized in the paraspinal, paravertebral regions. Retroperitoneal schwannomas are rare tumors that make up approximately 1 to 5% of all retroperitoneal lesions. The clinical picture of these tumors is due to the progressive growth of the neoplasm with the possible development of neurological symptoms. Clinical cases of malignant schwannomas with metastatic lesions of the lung, myocardium, perirenal adipose tissue on the right and left, and parapancreatic adipose tissue are described [8,10]. Articles have also been written about patients with malignant retroparietal schwannomas involving the inferior vena cava. Presented by the authors Stepanova Yu.A. and others, this clinical observation shows the difficulty in diagnosing retroperitoneal schwannoma. Careful preoperative diagnosis of the relationship and topography of the tumor

with all nearby organs and vessels according to various research methods allows the surgeon to determine the most optimal tactics for treating the patient [7]. Malignant schwannomas can occur in different localizations of the human body. For example, O.B. Laurent et al. described in detail the case of an intraoperative finding of a malignant schwannoma originating from the ureter [9]. This once again proves that this type of tumor is a diagnostic "mystery" for surgical doctors.

Conclusions. The above clinical observation demonstrates the difficulties in diagnosing malignant retroperitoneal schwannomas. It is of great importance in the preoperative period to determine the relationship of the tumor with all nearby organs and vessels according to various methods of radiation diagnostics. As well as a thorough histological and immunophenotypic study of the remote formation, it gives a chance to accurately determine the nature of the tumor and provides an opportunity to determine the correct further tactics for treating the patient.

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