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CLINICAL CASE: SURGICAL FINDING - MALIGNANT RETROPERITONEAL

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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 (TashGFRSNPMTSOiR) with a preliminary diagnosis of an ovarian tumor. Interrooperatively, a tumor of the retroperitoneal space with lesions of the obturator nerve was revealed. Subsequently, IHC analysis clarified the organ belonging and the histological type of the tumor, which turned out to be a malignant schwannoma. These data may help in the management of patients with this pathology and improve the diagnosis of patients with small pelvic tumors.

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

Introduction

Schwannomas, also known as neurylemmoms, are tumors consisting of well-differentiated Schwann cells originating from the glial cells of peripheral nerve membranes [1]. Most schwannomas are benign, and malignant ones, which are usually associated with von Recklinghausen's disease, are rare [2,3].

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Schwannomas usually occur in the head and neck, retroperitoneal space and limbs. The pelvic form is very rare, with a recorded frequency of 1-3% of all schwannomas [4,11]. Because there are no specific clinical or radiographic signs for pelvic schwanns, and they resemble a number of pelvic diseases, and can easily be misdiagnosed [5,6]. Surgical excision is both a diagnostic and therapeutic treatment for pelvic schwannome. In 50% of cases, schwannomas are combined with neurofibromatosis. The frequency of local relapses 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 with different morpho-functional properties that determine their clinical course [3]. The most common was the classification of L. Ackerman (1954), which the author developed on the basis of numerous works by the famous American pathologist Stout, published in 1943-1953. L. Ackerman believed that a satisfactory classification of retroperitoneal tumors at that time was hardly possible and called his separation of tumors of this localization conditional. All modern classifications are mostly based on the identification of the type of tissue from which the tumor is formed, that is, they are built on a histogenetic type. L. Ackerman also based his classification on his histogenetic traits, dividing 29 types of tumors into five classes. Malignant neurolemmama (malignant schwannoma) - a fairly accurate diagnosis and determination of the histogenesis of the tumor in these cases is associated with significant difficulties. Malignant neurolemmas are distinguished by the degree of maturity depending on the ratio of cellular and fibrous elements. These tumors, especially low-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 especially interesting for doctors of surgical specialties and pathologists.

Patient Z. 66 years old was admitted to the department of oncogynecology of the Tashkent city branch of the Republican Specialized Scientifically Practical Medical Center of Oncology and Radiology (TGF RSNPMTSOiR) on 14.11.22 with complaints of recurrent pain in the lower abdomen and lower back, frequent urination. From the anamnesis he is sick for 4 months. In July 2022, the patient began the above complaints, underwent ultrasound of the pelvic

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organs. On ultrasound, ovarian formations were detected. In the future, she turned to the polyclinic at the place of residence, examined by a gynecologist recommended additional examination. MRI of the small pelvis 09.11.22g: MRI - signs of uterine fibroids. Volumetric formation of the ovary. Inthe future, the patient was sent THF RSNPMCOiR, where she was examined by an oncogynecologist, additionally examined, Tumor markers from 05.11.22g: CA125 - 12.0 units / ml, HE4 68.7rM, ROMA index 13.3%. MRT (abdominal cavity, retroperitoneal space, small pelvis) from 09.11.2022: Conclusion: MRI signs of left-sided urethrohydronephrosis, uterine fibroids. Volumetric formation of the ovary.

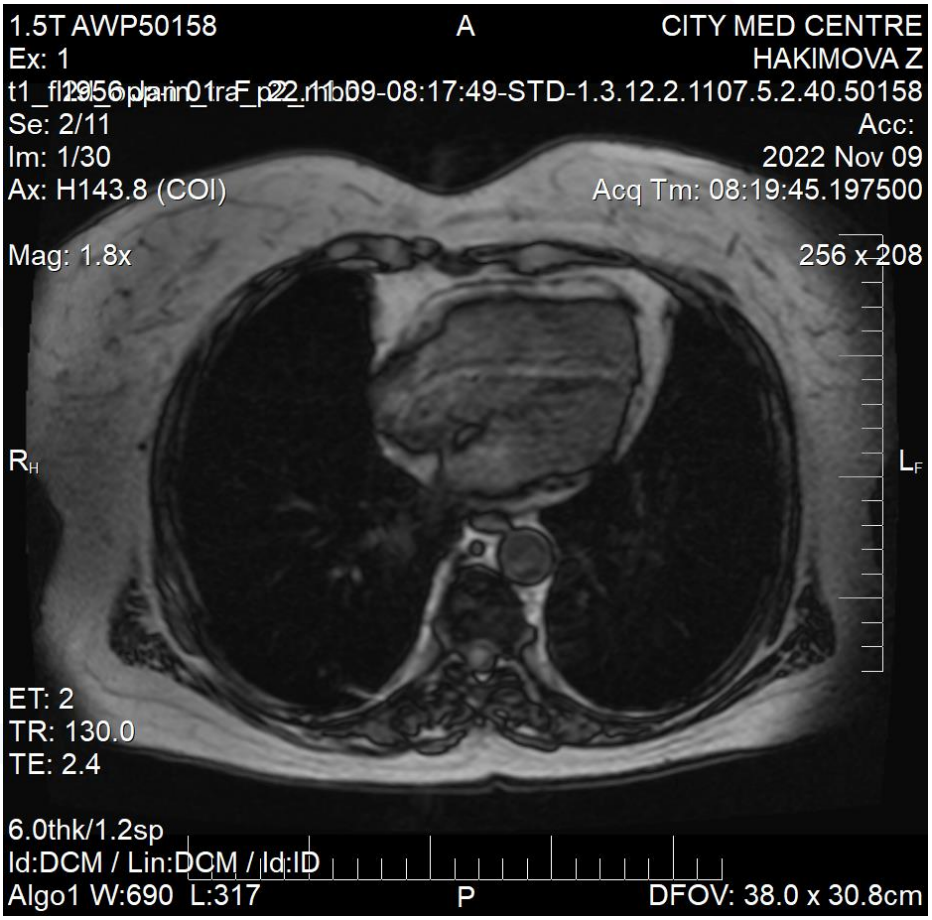

Figure 1. MRI examination of the small pelvis. MRI - signs of uterine fibroids. Volumetric formation of the ovary (horizontal incision).

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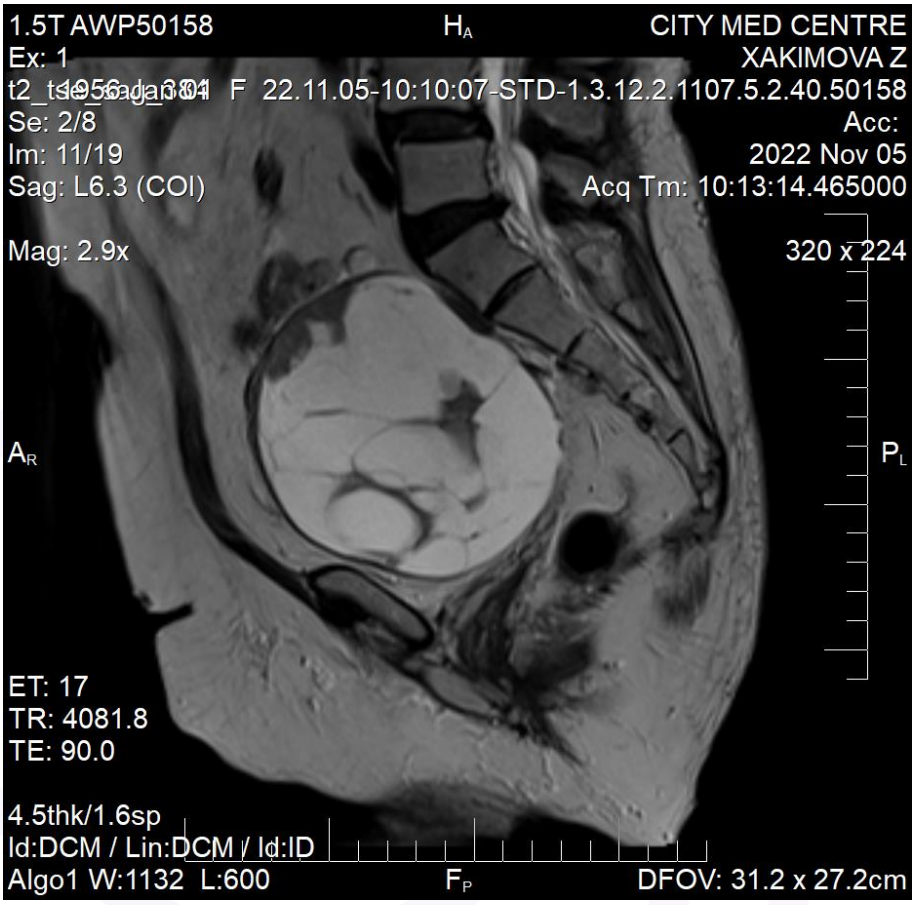

Figure 2. MRI examination of the small pelvis. MRI - signs of uterine fibroids. Volumetric formation of the ovary (sagittal incision).

In the future, the patient is diagnosed: Suspected tumor of the left ovary. Donkey: a symptom of pain. Urethrohydronephrosis on the left. The patient is hospitalized in the oncogynecology department for further examination and decision on further treatment tactics.

Upon admission, the general condition of the patient is satisfactory. The skin and visible mucous membranes of normal color. There is no hyperthermia. Breathing is vesicular, carried out in all departments, there is no wheezing pdd 16 in min. Heart tones are rhythmic, blood pressure 130/80 mm Hg. St. Pulse 78 in min. The tongue is clean, moist. The stomach is not swollen, on palpation it is soft, painless in all departments, participates in the act of breathing. Peristalsis is listened to. There are no peritoneal symptoms. The symptom of pricking is negative on both sides. The stool is adequate. Uricipinscania learned, in small portions, light urine.

According to the laboratory examination, an increase in ESR to 19 mm / h, a decrease in the level of hemoglobin to 96 g / l, a slight erythrocyte-, leukocyte-

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and proteinuria, a slight increase in the level of blood creatinine to 114 μmol / l were revealed.

With ultrasound: in the abdominal cavity, retroperitoneal space, as well as in the pelvis, free fluid and delimited liquid 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 one is 114x56 mm in size, the parenchyma is uniformly thinned to 5 mm, increased echogenicity, pronounced calicopieloureterectasia (pelvis 56 mm, calyx up to 53 mm); In the pelvis, more on the left in the projection of the left ovary, the formation of an irregular shape is determined, with dimensions of 181.2x99.8x13.7 mm. The formation is heterogeneous in structure, mainly solid (heterogeneously reduced echogenicity) with the presence of cystic cavities of various shapes and sizes (the contents of some cavities are diffusely heterogeneous). The uterus in RFV, measuring 65x74x56 mm, myometrium of homogeneous structuresy. Endometrium - 3 mm. Right ovary size 30x26 mm, homogeneous structure. Conclusions: Tumor of the left ovary. Ureterohydronephrosis on the left I degree.

Radiography of the chest organs: fresh focal and infiltrative shadows were not detected.

EhoKG: The general and regional contractility of the left ventricle is not impaired. The valves and septa of the heart are intact. The cavities of the left ventricle are not dilated. The pericardium is not dilated. Tachyarrhythmia. Dandastolic dysfunction of the left ventricle type 1 typeA.

Doppler of the lower tothe numbness. Conclusion: The main arteries of the vessels of the lower extremities are passable. Hemodynamic-significant stenosis was not revealed.

EGDFS: Insufficiency of the ring of cardia, without appendages of reflux esophagitis. Possible axial hernia of the esophageal opening of the diaphragm. Superficial gastritis. After an ulcerative linear scar along the anterior wall of the duodenal bulb. Erosive bulbit.

Cytology No. 105886/22: posterior vault puncture: Cytological picture with suspected cell atypization. For the differentiation and final diagnosis of neoplasm, additional studies are needed.

St. Genitalis: NGOs are developed correctly, female-type hair growth. In the mirrors: the cervix is retracted to the right, the discharge is light. P.V: The body

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of the uterus and appendages in a single conglomerate, palpated formations measuring 15 cm in diameter, limited mobility. The parameters are free.

According to the examination, the patient was diagnosed with a preoperative diagnosis: Main: Tumor of the left ovary. Complications: Ureterohydronephrosis on the left of the 1st degree. A symptom of pain. Concomitant: Chronic bronchitis. After ulcerative duodenal scar. Catarrhal gastritis.

23.11.2022 the operation was performed: LAPAROTOMY, REVISION, EXTIRPATION OF THE UTERUS WITH APPENDAGES, REMOVAL OF A TUMOR OF THE RETROPERITONEAL PROSTRATION.

Course of the operation: Under epidural anesthesia, a lower median laparotomy of 18 cm in length was performed, the thickness of the subcutaneous fat is up to 5 cm. When revising: 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 arteries, ureter, and the posterior wall of the bladder.

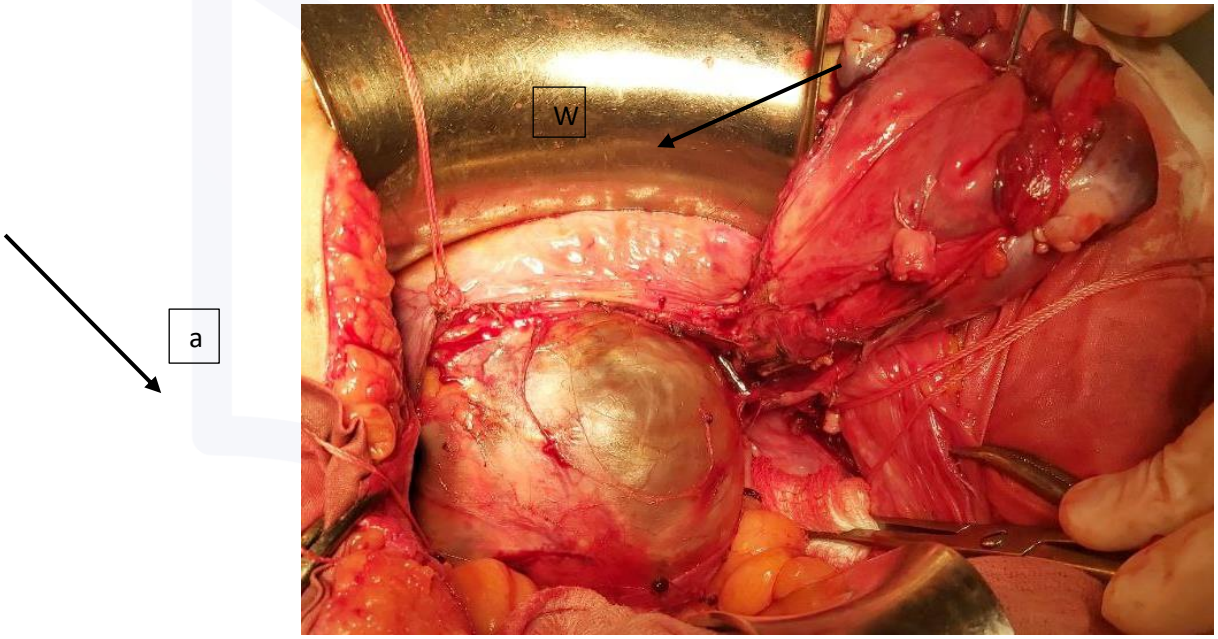

Figure 3. Stages of the operation: a - cystic part of the tumor, b - uterus with appendages

The body of the uterus is enlarged to 7 weeks of pregnancy, densely - elastic consistency, deviated to the right due to a tumor (Fig. 3). Cervix measuring 2.0x4.0 cm, hypotrophied. The pipes on both sides are unchanged. The ovaries on both sides are cystically altered, measuring 4x3.5 cm. The liver and other

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organs of the abdominal cavity - without visible pathology. The round ligaments on both sides and the funoxtase ligaments on the left are clamped, crossed, stitched and doped. The vesico-uterine and rectal-uterine folds are acutely deflated. On the cervical vessels on both sides clamps are applied, crossed, stitched and ligated. Extirpation of the uterus with appendages was performed. The stump of the vagina is sutured tightly. Hemostasis.

Mobilization of the retroperitoneal tumor on the left began, with technical difficulties the separation of large vessels from the tumor, especially the external iliac vein and artery, was carried out. The tumor grows into the mesentery of the sigmoid colon, the adhesions are dissected acutely, atraumatic gray-serous sutures are applied to the deserosed areas of the intestine for a length of 8 cm. Further revision revealed that the tumor grows into the internal iliac vein and artery (Fig. 4), taking into account the above, the internal iliac artery and vein are crossed, stitched and doped.

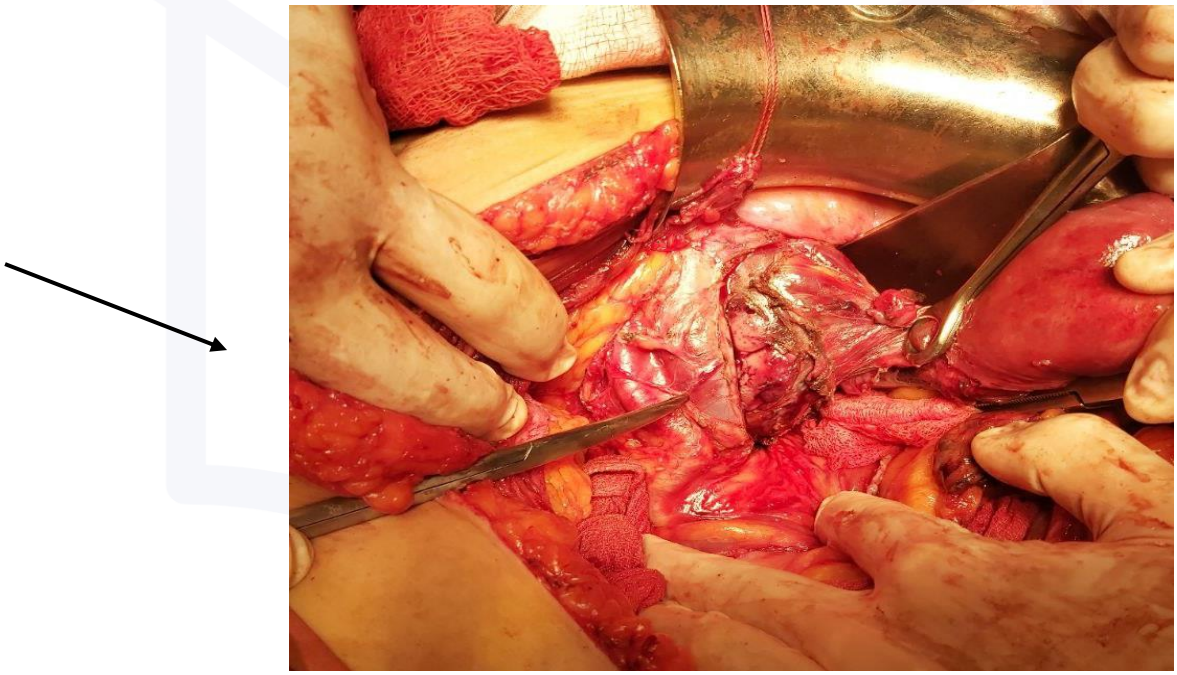

Figure 4. Stages of surgery: detection of tumor germination in the internal iliac vein and artery.

The distal part of the ureter on the left was mobilized, while it was revealed that there is an expansion of the ureter. Part of the tumor was sent for express histological analysis. Answer to urgent histological analysis No. 11589: alveolar rhabdomyosarcoma.

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Upon further revision, it was found that the obturator nerve passes inside the tumor (Fig. 5). Relatives of the patient's preoperative room were called, a conversation was held about the response of urgent histology, about the germination of the tumor into the vessels and nerve innervating the left leg, possible complications of the disease, consent was taken to remove the nerve. Given the germination of the tumor into the obturator nerve and the defeat of this site inside the tumor, it was interoperationally decided to resect part of n.obturatorius with the tumor from the principle of radicalism.

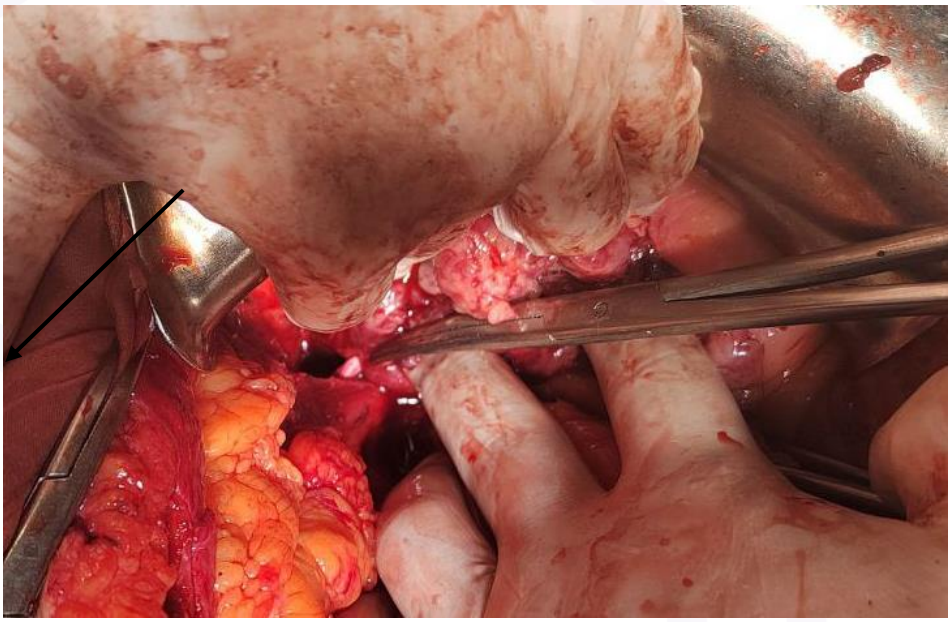

Figure 5. Stages of the operation: an obturator nerve passes inside the tumor.

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

Macropreparation: The body of the uterus is enlarged to 7 weeks of pregnancy, dense-elastic consistency, diffusely hypertrophied on the myometrium incision, the endometrium is hyperplastic. The cervix is 3.0x3.5 cm in size, densely elastic consistency on the incision of multiple Ov.Nabothii, the c / canal is smooth. The pipes on both sides are unchanged. The ovaries on both sides are cystically altered, measuring 4x3.5 cm. A tumor of the retroperitoneal

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space measuring 15x20x25 cm, cystic - solid structure, with part of the obturator nerve, a solid part of the tumor is loose, grayish in color, resembling "fish meat".

The course of the postoperative period is smooth. The stitches were removed on the 10th day. The urethral catheter was removed on the 10th day after the operation, independent 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 section 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. 6). A focus of necrosis is noted. Mitotic activity is 1-2 / per 1 field of view. The tumor grows into the surrounding fibrous tissue and one reactive node.

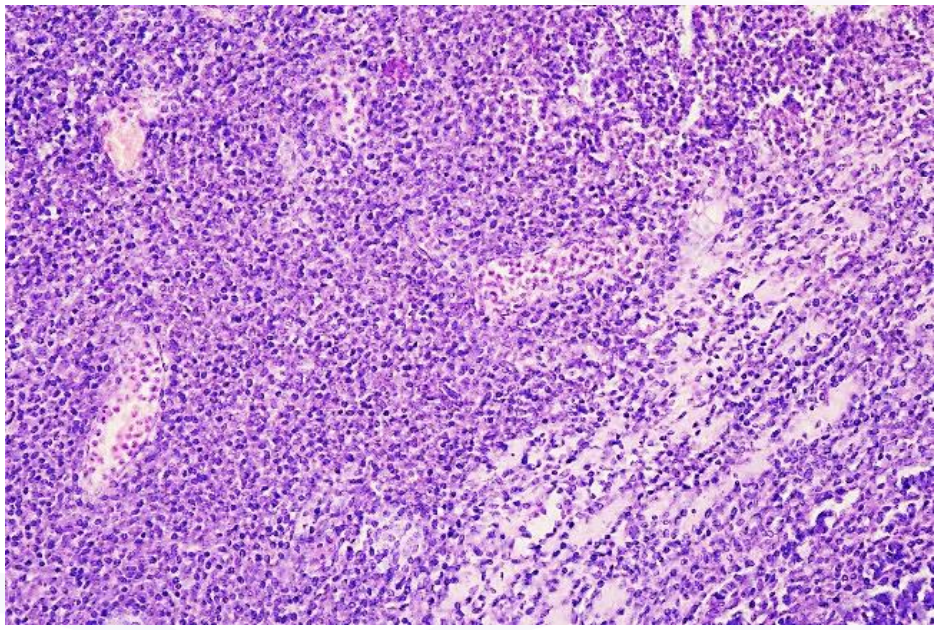

Figure 6. Microscopic picture of intraoperative material.

In the myxoid stroma with foci of hyalinosis, hypercellular areas are visible, consisting of malignant cells from a rounded to spindle-shaped structure. Cells with moderate polymorphism, the cytoplasm is scanty, nuclei with visible vesicular nuclei.

With a planned histological examination, the morphological picture of a malignant neoplasm had similar signs with such tumors as rhabdomyosarcoma, leiomyosarcoma, extraintestinal GIST, synovial sarcoma, myeloid sarcoma and malignant schwannoma. Neoplasia resembles a biphasic

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tumor consisting of alternating foci of various structures. There are layers of closely spaced spindle-shaped structures, as well as small rounded, blue cells with signs of immature differentiation (Fig. 7, 8) .

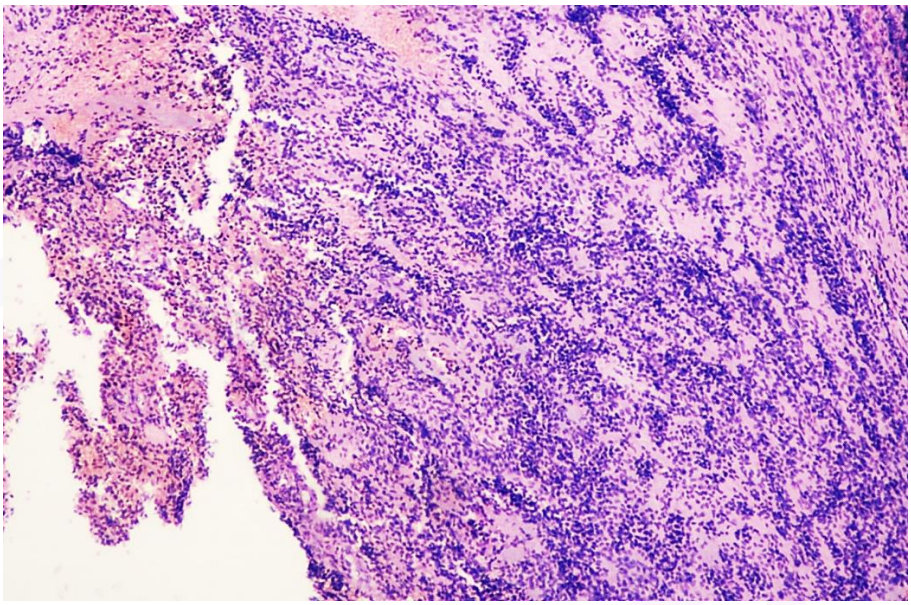
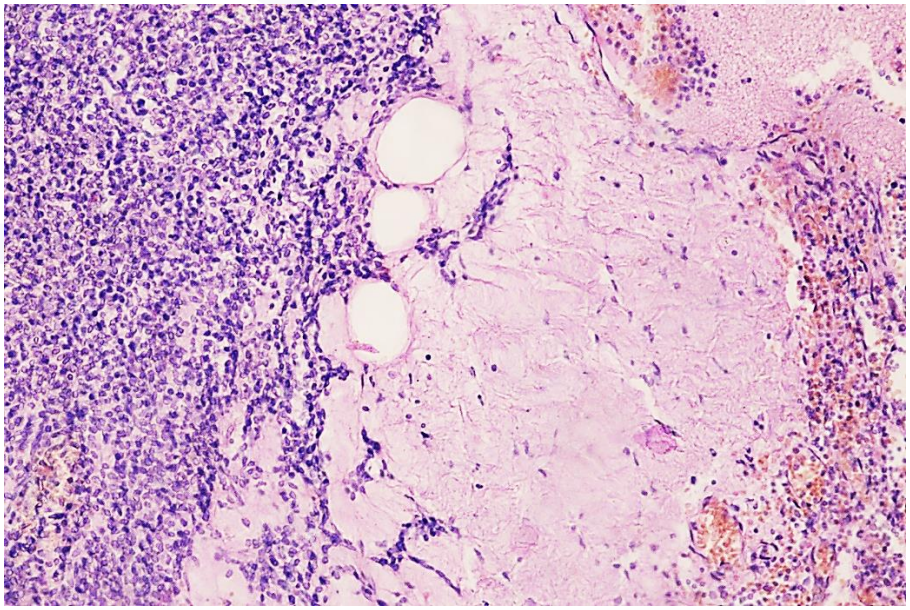


Figure 7. Morphostructure of the removed tumor. Foci of hyalinized stroma with oval and rounded cells building psedvomomary structures (ca. GE. X100) are noted.



Picunoc 8. Morphological picture of a low-differentiated neoplasm. There are foci of proliferation of small rounded cells with vesicular nuclei. (Ok.GE. x100).

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To clarify the diagnosis, an immunohistochemical study was performed. During immunohistochemical profiling of the tumor, Myogenin, MyoD1 and Desmin were negative and the initial diagnosis of rhabdomyosarcoma was removed. After that, an IHC marker panel was created, including PanCK, CD117, DOG1, CD34, SMA, BCL2, MPO, TdT, CD99 and S100. To our surprise, PanCK, CD117, TdT, CD99 stained minor areas in the tumor tissue. Since CD34 and DOG1 were negative, GIST was not confirmed. Despite the focal positivity of TdT and CD99, the MPO was negative, and therefore the diagnosis of myeloid sarcoma was also excluded. Biphasic synovial sarcoma and malignant schwannoma remained for differential diagnosis. Given the diffuse and strong expression of S-100 and the negativity of BCL2, synovial sarcoma was also excluded. Additional markers were placed to exclude melanoma. HMB45, MelanA and SOX10 were negative. Immunoprofiling of the resected tumor corresponded to a low-differentiated malignant schwannoma (Fig. 9).

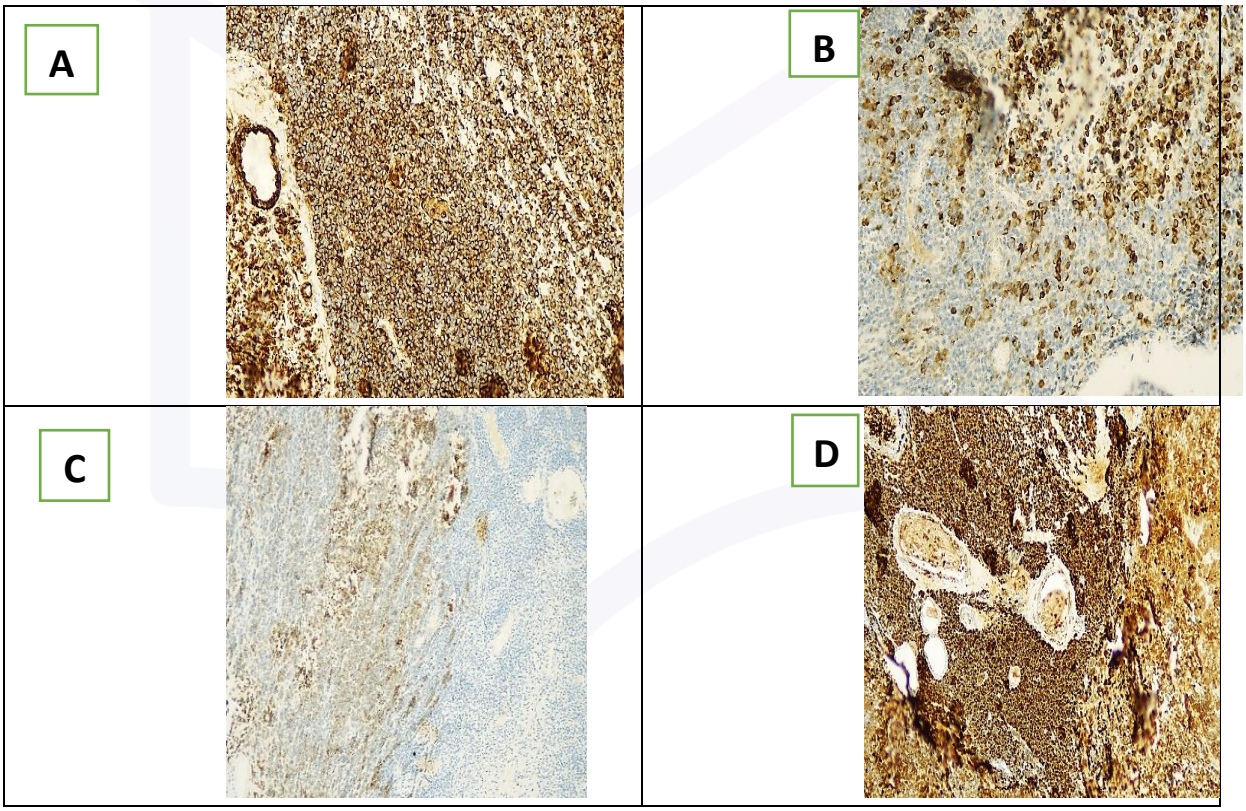

Figure 9. IGC picture of low-grade malignant schwannoma.

A. Diffuse positive expression vimentin (IHC staining, x100). B. Focal positive expression to PanCK (IHC staining, x100). C. Focal positive expression to CD117 (IHC staining, x100). D. Diffuse positive reaction to S100 (IHC staining, x100).

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Discussions. Malignant schwannoma or the so-called malignant neurolemma, develops from the Schwann nerve sheath and accounts for less than 5% of all malignant soft tissue formations. By localization, malignant neurolemmoma most often affects the nerve plexuses of the upper half of the trunk, neck, upper extremities, small pelvis, less often localized in the paraspinal, paravertebral regions. Retroperitoneal schwannomas are rare tumors that range from about 1 to 1 to 5% of all retroperitoneal formations. 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, paranephral fatty tissue on the right and left, parapancreatic fatty tissue [8,10] are described. Articles have also been written about patients with malignant schwannomas of retroparietal location with involvement of the inferior vena cava in the process. Presented by the authors Stepanova Yu.A. et al., this clinical observation shows the difficulties in diagnosing schwannoma of the retroperitoneal space. Thorough 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 and co-authors described in detail the case of an intraoperative finding of an ill-quality schwannoma emanating 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 schwannomas of the retroperitoneal space. 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 diagnosis. As well as a thorough histological and immunophenotypic examination of remote formation gives a chance to accurately determine the nature of the tumor and provides an opportunity to determine the nature of the tumor determine the correct further tactics of treating the patient.

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