

Case Report

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Leiomyosarcoma in A Patient with Accessory Kidney - Clinical Problems, Imaging and Treatment

Emil Enchev*, E. Dimitrov, G. Minkov, St. Nikolov, I. Ovcharov, Y. Yovchev

Clinic of Surgical Diseases, University Hospital, Stara Zagora, Bulgaria

*Corresponding author: Emil Enchev, Clinic of Surgical Diseases, UMHAT "Prof. Dr. St. Kirkovich" AD, Stara Zagora, Bulgaria. Tel: +359894845263; +359884099549; Email: emo_19_89@abv.bg

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Abstract

Primary retroperitoneal tumors are of interest to several surgical specialties, including surgeons, urologists and gynecologists. We describe a rare clinical case of retroperitoneal leiomyosarcoma comprising a third accessory kidney. Physical examination of the abdomen and the left hypochondrium established a tight-elastic formation. The subsequent instrumental tests revealed the presence of a third kidney in the relevant anatomical area and contributed to the preoperative verification of the palpable formation. In contrast, we did not detect laboratory abnormalities. After preoperative preparation, radical extirpation of the tumor formation was performed together with the additional kidney involved. In conclusion, the main problem remains the late diagnosis, which in most cases is the cause of the poor late results in this type of tumors.

Keywords: Clinic, Diagnosis; Retroperitoneal Leiomyosarcoma; Treatment

Introduction

Although primary retroperitoneal tumors are relatively rare (0.2%) [1,2], they still present a challenge in terms of diagnosis, behavior and treatment. Their localization, however, also creates confounding diagnostic and healing difficulties. For the first time, the term retroperitoneal tumor was introduced in 1842 by Lobstein [3], and even today there is no noticeable progress in the disease. In spite of technical progress and the success of immunology and genetics, surgical treatment remains at the heart of the healing process, resulting in a significant improvement in patient life. Interferences in the retroperitoneal space require surgical knowledge in the areas of gastrointestinal, urological and vascular reconstruction techniques [1,4-6] Although primary retroperitoneal tumors represent a small percentage of the total number of tumors - 0.2% [1,2], they mostly affect the age limit between 40 and 70 years. An exception is also made for those found in children or in adults. The retroperitoneal space contains a number of loose tissues and specific anatomical elements, which is why they are diagnosed histologically and there are different types of retroperitoneal tumors.

In adults, lymphomas and sarcomas are the most common, and neuroblastoma - in children [1,3] Renal sarcomas are diagnosed in 0.8% to 2.7% of all renal tumors, and 50-60% after histological examination reveals the presence of leiomyosarcoma. Nevertheless, his aetiology remains unclear for now.

Clinical case

The clinical case we want to present concerns a female patient at the age of 75. The patient enters the clinic on the occasion of an echographically established formation in the retroperitoneal space. Anamnestic complaints are expressed in diffuse pain, radial to the front of the abdomen and intermittent gastrointestinal disorder.

Laboratory results

The blood counts and biochemical measurements performed did not detect deviations from the reference values.

Instrumental research

An echography study found a formation in the left retroperitoneal space. The CT showed 40/20 cm formation and abnormality in the urinary-genital system - an additional kidney covered by the formation (Figure 1-4).

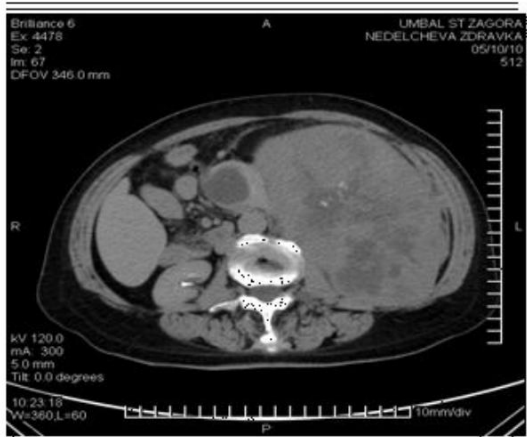


Figure 1: Sagittal cut - CT find.



Figure 2: Transversal cut of the retroperitoneal tumor.

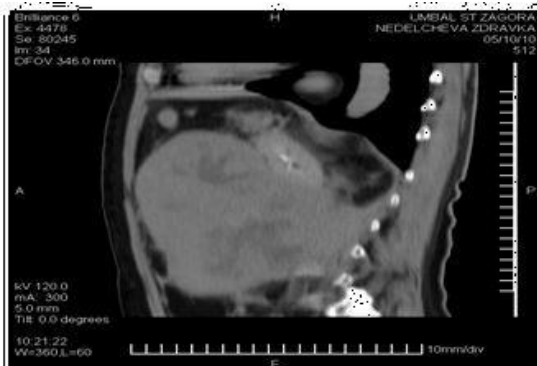


Figure 3: CT image of a formation engaging the left retroperitoneal space.

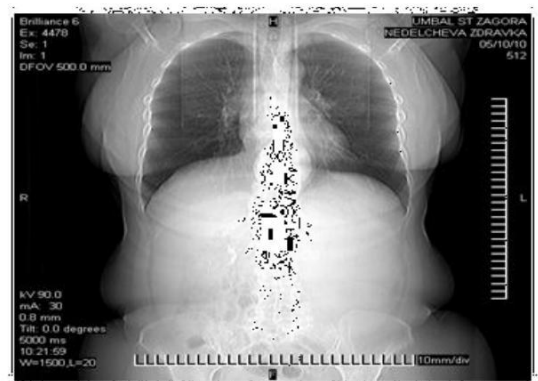


Figure 4: Venous urography - third additive kidney.

Treatment

The patient underwent surgical intervention under general anesthesia. The wide transabdominal access provided an exploitation of the abdominal cavity and retroperitoneum. Intraoperatively, we detected infiltration of the lower left kidney. We managed to re-sect the tumor radically (R0). We have subsequently documented the presence of the third free kidney located in the tumor formation (Figures 5A-D, 6).

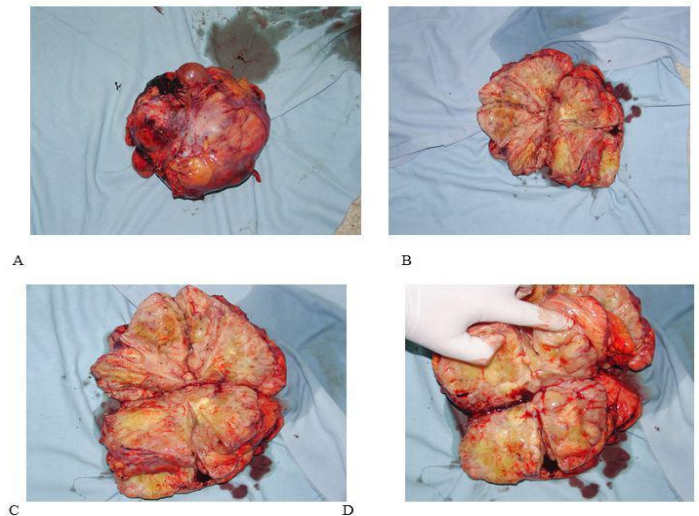


Figure 5A-D: Macroscopic appearance of the retroperitoneal-located formation.

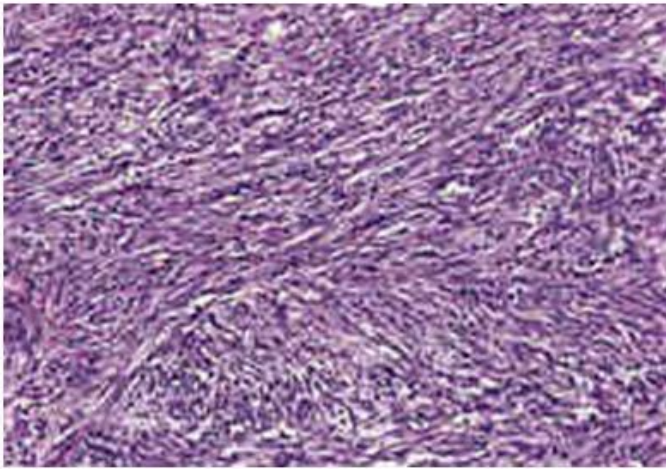


Figure 6: Histological picture of leiomyosarcoma.

The histological study performed with Vimetin - / ++ /; Desmin - / ++ /; SMA - / ++ / detected moderately differentiated leiomyosarcoma. According to TNM, for the sarcomas, the process was T2bN0M0 - stage IIB.

Discussion

There are no clear clinical symptoms of retroperitoneal tumors and additional clinical signs and symptoms are often result of involved adjacent organs in the process. In a number of cases, this causes a misdiagnosis. The imaging methods used to diagnose retroperitoneal tumors such as ultrasound, venous urography, CT, MRI, PET/CT are characterized by their tricks and disadvantages. Ultrasound, as a diagnostic method, provides an approximation of the tumor. The advantage of the method is its easy accessibility and repeatability as well as lack of exposure. Venous urography is extremely useful for the study of patients who have compression of the ureters or affecting the kidneys themselves. It can detect both the presence of hydronephrosis and diagnose an urographic "Mute kidney". In our case, the method detected the presence of a third kidney. At present, information obtained from CT, MRI is crucial for the upcoming surgical treatment. These methods make it possible to determine the exact size, content and invasion of the process. Another advantage of the methods is that in some cases the histological type of the tumor (angioliroma) can be determined with their help [4]. Not least, Positron Emission Tomography (PET) is very useful for detecting and treating relapses after a radical surgery.

According to other authors, and in our clinical case, laboratory tests proved to be extrinsic. Sometimes deviations can be

observed and found in biochemical tests, but this change is related to invasion of the process in a neighboring organ. Thus, elevation of bilirubin is most often found after biliary tract involvement [1]. There are no specific tumor markers yet. In some histological types of tumors there are increases in some antigens (CA19-9, CEA, alpha-fetoprotein, etc.) or other substances such as interleukins, as well as a decrease in the number of neutrophil lymphocytes or T-lymphocyte variations. In the same way, no changes in the genetic studies under way have been identified [7,8]. The treatment of retroperitoneal tumors is predominantly surgical, aiming at removing the tumor at the oncologically radical boundaries. When this is not possible, partial cuts or biopsies help ensure that the diagnosis is correct. Primary, unlike metastatic retroperitoneal tumors, are few chemically and radiosensitive. Radiotherapy should be used for size reduction or as a method of additional postoperative control.

The volume of operative intervention determines post-operative evolution of leiomyosarcoma. If the tumor is completely eliminated, which we have managed to achieve, the risk of further development of the disease remains minimal [1,3,4]. Even today, retroperitoneal tumors are a special feature in oncology. Located deep in the retroperitoneum, they often grow into adjacent structures - so removal of tumors is sometimes accompanied by entero-tomy, splenectomy, nephrectomy, colectomy or partial resection of vessels. That's why this pathology remains a real challenge for the surgeon [2,4,6], but at the same time they are no longer "Terra Incognita".

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