Case Reports
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SPONTANEOUS HEMOPERITONEUM SECONDARY TO RETROPERITONEAL TUMOR RUPTURE

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INTRODUCTION

Spontaneous hemoperitoneum is the presence of blood in peritoneal cavity without antecedent trauma. The clinical features are abdominal pain, sometimes severe, bloating, decreased hematocrit and occasionally signs of hypovolemic shock. The most common causes are (1, 2):

- Iatrogenic: surgery, interventional procedures, anticoagulant therapy, chemotherapy treatments.
- Tumors rupture: hypervascular tumors, like liver cancer (most common).
- Pathology vascular (aneurysms or pseudoaneurysms)
- Blood dyscrasias: hemophilia, polycythemia vera.
- Rupture of Spleen: infections, cysts, metabolic diseases.
- Gynecologic Pathology: ruptured ectopic pregnancy, ruptured ovarian cyst, HELLP syndrome.

Resumen.- OBJETIVO: Describimos un caso de tumor retroperitoneal tipo GIST con rotura espontánea a cavidad abdominal, ocasionando un cuadro de abdomen agudo secundario a hemoperitoneo.

MÉTODO Y RESULTADOS: Varón de 84 años que acude a servicio de Urgencias de nuestro hospital por cuadro sincopal, con dolor abdominal difuso y cortejo vegetativo acompañante. Tras la realización de diferentes pruebas complementarias se objetiva una masa retroperitoneal dependiente de riñón izquierdo de unos 19 cm con sangrado activo y hemoperitoneo secundario, por lo que se realiza una nefrectomía radical izquierda con resultado anatomo-patológico de tumor estromal gastrointestinal dependiente de la cápsula renal.

CONCLUSIONES: El hemoperitoneo espontáneo es una entidad poco frecuente y de una etiología variada estando descrita en muy raras ocasiones en tumores retroperitoneales.

Palabras clave: GIST. Retroperitoneo. Hemoperitoneo.

Summary.- OBJECTIVE: To report a case of GIST type retroperitoneal tumor with spontaneous rupture to the abdominal cavity causing acute abdomen secondary to hemoperitoneum.

METHODS/RESULTS: We report the case of an 84 year-old man with history of BPH and chronic atrial fibrillation. He presented to the Emergency Department with diffuse abdominal pain, syncope and accompanying vegetative symptoms. Diagnostic work up showed a 19 cm retroperitoneal mass dependent of the left kidney with active bleeding and secondary hemoperitoneum. Left radical nephrectomy was performed with pathology report of gastrointestinal stromal tumor attached to the renal capsule.

CONCLUSIONS: Spontaneous hemoperitoneum is a rare entity and it has various etiologies. It is rarely described in retroperitoneal tumors.

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Pathology of the digestive tract, peptic ulcer bleeding, tumor.

It has also been described in spontaneous ruptures of lymphatic metastasis of testicular germ cell tumors and retroperitoneal leiomyosarcoma (2, 3).

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors in gastrointestinal tract, defined by immunohistochemical studies performed by Mazur and Clarks, and CD 177 positive mesenchymal tumors, spindle cells (77%), epithelioid cells (8 %) or mixed cells of both spindle and epithelioid types (pleomorphic) (15%), primary gastrointestinal tract, omentum, mesentery and retroperitoneo (4).

It’s more prevalent between 40 and 60 years of age. In United States, between 3,000 and 5,000 people each year develop GIST (5).

The origin of this neoplasm are the interstitial cells of Cajal, the “pacemaker cells of the gut” that send nerve signals to propel food along its course through the system via muscle contractions (peristalsis), showing mutations in the Kit proto-oncogene.

The most frequent site for GISTs is the stomach (about 55%), followed by the duodenum and small intestine (about 30%), esophagus (about 5%), rectum (about 5%), colon (about 2%), and rare other locations.

In extraintestinal sites include the mesentery, omentum and retroperitoneum, with a low frequency, less than 5%, defined as extragastrointestinal stromal tumors (EGIST) (6,7).

CASE REPORT

We report the case of a 84 year old man with a history of BPH and chronic atrial fibrillation. He attended to the A&E department with diffuse abdominal pain, syncope and accompanying vegetative.

In the physical examination revealed a BP of 90/60, Fc 120, pale skin and mucous membranes, with a good level of consciousness. Physical examination showed a great abdominal mass in the middle left. Blood test showed a hemoglobin of 8 g / dl, hematocrit of 25% with leukocyte count and normal coagulation. Creatinine was 1.2mg/dl.

CT scan showed normal-sized right kidney and left kidney unstructured with anterolateral displacement by vascularized retroperitoneal mass with areas of low attenuation suggestive of necrosis(dimensions of 15.8 x 18 x 6.5 cm), which depended of left kidney.

Figure 1. CT scan with tumor.  
Figure 2. CT scan with tumor.
Also showed a cortical lesion in middle third of left kidney with irregular morphology, hypodense, with parietal calcifications, suggestive of cystic (Figure 1 and 2), retroperitoneal and pelvic lymph nodes, and moderate volume of free fluid (45 HU) suggestive of hemoperitoneum, located in pelvis and perihepatic.

Patient presented active bleeding by the mass and we decided transperitoneal radical nephrectomy urgent through an anterior subcostal, demonstrating active bleeding in surgery room by lesion in peritoneum about 3 cm. (Figure 3).

The Pathological study revealed the existence of gastrointestinal stromal tumor (retroperitoneal) (EGIST) of about 29 cm in size, spindle cell carcinoma, infiltrating kidney. Mitotic rate of 42/50 hpf. Presence of necrosis (<50%). Renal parenchyma without changes (Figure 4).

After surgery, the patient presented good response and at present, the patient is asymptomatic.

**DISCUSSION**

EGIST are extremely rare tumors, with few studies and case reports in the literature (6), with incidence estimated at 20 cases per million habitants, more usual in males around 50-70 years.

In radiology studies it’s present as a great solid mass (3-10cm), hypervascular and variable density on CT with IV contrast. Small lesions are, usually homogeneous, while large are heterogeneous, with bleeding and necrosis (1).

Pathological diagnosis include structural features such as evidence of hyaline stroma with abundant collagen fibers, a lymphocytic infiltrate and the presence of myxoid stroma.

The clinical is variable, from paintings at the occlusive jejunoileal or colonic, gastrointestinal bleeding in the stomach or mass with or without compression of structures in the retroperitoneal. In our case, patient had a secondary hemoperitoneum, extremely rare, setting the literature less than 40% of GISTs, without date on the frequency in the extragastrointestinal stromal tumors, by limited number of cases, constituting all of them a surgical emergency.

The liver is the most common site of metastasis of these tumors, with an incidence around 55-72%.

For their diagnosis are useful ultrasound, CT and MRI. Another option is a biopsy of the mass for establishing the diagnosis, although there is a high risk of bleeding and dissemination.

Prognostic factors that allow an approximation of the biological activity and aggressiveness of these tumors have been studied by different authors such as Fletcher, who in 2002 proposed the recurrence risk groups based on the mitotic rate and tumor size 8. Later, Miettinen modified risk index by adding the location of the tumor in gastrointestinal (9) tract, although in the case of EGIST, presence of necrosis (which correlates with large tumor size), high cellularity and mitotic rate show a worse prognosis (Reith et al) (10). Also, there are genetic markers with prognostic value, such as gain and loss of genetic material, deletions or gene overexpression VIL2 or FAK as well as mutations of c-Kit, although today there aren’t conclusive data (10).

For the differential diagnosis should be considered tumors with expression of CD34 or c-Kit, such as fibrohistiocytic, or leiomyosarcoma (9,10).
At recurrence of leiomyosarcoma diagnosed and previously operated immunohistochemistry should be performed to diagnose a GIST.

As treatment modalities, surgery is the only potentially curative treatment for primary gastrointestinal stromal tumor, if acceptable morbidity and it’s made with radical character, with complete removal of the pseudocapsule, and with good margin of safety (9).

Another treatment option as adjuvant treatment with imatinib mesylate after surgery in order to cause the regression of the interstitial cells, which lose their activity c-Kit, although it should be considered experimental, following the recommendations of the Spanish Group of sarcomas, in the absence of randomized studies to support its use.

Another possible treatment in cases of metastatic GIST is the use of antiangiogenic therapy based on Sunitinib as second-line treatment, especially in those cases refractory to therapy Imatinib (10).

CONCLUSIONS

GIST tumors are extremely rare tumors in the retroperitoneum, so the case report is doubly interesting, by the presentation and histological type.

REFERENCES AND RECOMMENDED READINGS (*of special interest, **of outstanding interest)