A rare case of pelvic solitary fibrous tumor

Clinical History:

A 61-year-old man presented recurring episodes of wake-up confusion and hallucinations. These symptoms normalized during the day, the patient had no recollection of the episodes. He had no relevant medical background. Neurological examination and electroencephalogram were normal at admission. During the hospitalization, nocturnal hypoglycemia of unknown origin was observed.

Imaging Findings:

An abdominal CT without and with contrast media was made to exclude a pancreatic lesion. It showed a well delimited pelvic mass, retroperitoneal in left paramedian position. Without contrast media, the lesion showed a mesenchymal density (mean of 45 Hounsfield units) without any calcifications and a weak heterogeneity. The bladder and the sigmoid were shifted to the right. In arterial phase, this mass showed a direct vascularization by a branch of the left internal iliac artery. In the portal phase, it showed an important and heterogeneous peripheral enhancement.

There was no fat infiltration and no lymphadenopathies around it. Angiography confirmed the hypervascular nature of the tumour, by showing a central vascular pedicle taking its origin from the left internal iliac artery with an arborizing pattern of vessels. After 4 years, a follow-up CT showed a new mass in the peritoneum in the left hypochondria with the same characteristics.

Discussion:

Solitary fibrous tumours (SFTs) are rare mesenchymal neoplasms and account for less than 2% of all soft-tissue tumours.[1] They belong to a unique group of mesenchymal neoplasms of fibroblastic or myofibroblastic origin.[2] Initially, they were thought to be confined to the pleural serosal surfaces (due to a putative mesothelial or submesothelial origin) but it is now established that SFTs are ubiquitous neoplasms and extrapleural location is considered to be more common.[2, 3] They can appear in various locations such as the head and neck, abdomen, pelvis, and also in the periosteum and somatic soft tissues.[2]
It occurs commonly in the 5th decade of life without gender predilection.[4]
They manifest as a slow-growing mass and most of them are asymptomatic.[2] The most common symptoms are pain, palpable mass or symptoms due to pressure effect (urinary or bowel obstruction).[2] Hypoglycemia is a rare paraneoplastic manifestation of SFT and is seen in less than 5% of cases; it is referred to as Doege-Potter syndrome.[2] This hypoglycemia is related to an excessive production of insulin-like growth factor 2 by the tumour.[5] This results in stimulation of the insulin receptors and increased glucose utilization.

On a contrast media enhanced CT, SFT appears as a well-defined mass that may exert pressure effects on neighbouring organs. It presents an intense heterogeneous enhancement that persists on delayed phase. Sometimes it may be seen areas of central low attenuation, representing necrosis, haemorrhage or cystic changes. Calcifications are rare and usually seen in large tumours. [3] MRI was not performed in our case, however, SFT typically presents an intermediate T1 signal intensity and heterogeneous low T2 signal intensity with flow voids with intense arterial enhancement observed after administration of gadolinium. Peripheral serpentine vessels adjacent to the tumour can reinforce the diagnosis. [2]

At angiography, SFTs typically appear as a highly vascular mass with arborizing vessels arising from a vascular pedicle. Dilated arteries and early visualization of veins may also be seen.[2] SFTs are usually benign but can present an aggressive behaviour in 10-15% of cases.[2]
The treatment of choice is the complete surgical resection.[6] Preoperative embolization can reduce intraoperative haemorrhage.[2]

Long-term follow-up is recommended.[7]

Prospective imaging diagnosis of SFT is unlikely. Nevertheless, unexplained hypoglycemia in addition to the finding of a hypervascular mass should lead to the diagnosis of Doege-Potter syndrome, which is a rare paraneoplastic manifestation of SFTs.

**Differential Diagnosis List:** Pelvic solitary fibrous tumor confirmed at pathology after complete resection, Pelvic hypervascular tumours, Mesenteric fibromatosis, Metastatic carcinoid tumour, Inflammatory pseudotumo, Lymphoma

**Final Diagnosis:** Pelvic solitary fibrous tumor confirmed at pathology after complete resection

**References:**

**Description:** Pelvic mass (star) measuring 11 x 14 x 11 cm, situated in the retroperitoneal to the left of the bladder. The bladder and the sigmoid are shifted to the right (arrows). **Origin:** ? Department of Radiology, CHU Ambroise Paré, Mons, Belgium, 2020
Figure 2

Description: It showed a direct vascularization of the mass by a branch of the left internal iliac artery (arrow). Origin: ? Department of Radiology, CHU Ambroise Paré, Mons, Belgium, 2020
Description: Important and heterogeneous peripheral enhancement of the mass.

Origin: ?

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Figure 4

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Figure 5

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