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PANCREATIC METASTASES FROM A RECURRENT MENINGEAL SOLITARY FIBROUS TUMOR: ABOUT A CASE

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Abstract

Solitary fibrous tumor (formerly hemangiopericytoma) is a rare, aggressive and highly metastatic tumor of the soft tissues and meninges. The majority of metastases occur in bone, lung and liver, and rarely in other organs. Pancreatic metastases are extremely rare. We report here a rare case of recurrent meningeal solitary fibrous tumor with pancreatic metastases in a woman who had a surgical resection 18 years earlier, and describe the CT and MRI imaging features.

Keywords: Pancreas; Solitary Fibrous Tumor; Hemangiopericytoma; Metastasis; Meningeal, CT, IRM.

INTRODUCTION

Hemangiopericytoma (HMP) is a rare mesenchymal tumor derived from Zimmerman pericytes [1].

Theoretically, the tumor can arise anywhere there are capillary beds, most commonly in the soft tissues of the lower extremities, pelvis, or retroperitoneum. Regardless of its location, it is an aggressive tumor with a high rate of local recurrence and metastases. In the central nervous system, as of the 5th edition (2021) of the WHO classification of CNS tumors, the term hemangiopericytoma has been removed and is now included as a continuum of solitary fibrous tumor (SFT). [2]. Intracranial TFS are rare and correspond to 2 to 4% of meningeal tumors and less than 1% of all central nervous system tumors. The most common metastatic locations are bone, lung or liver. Other metastatic locations are rarer, such as the kidney, pancreas, adrenals, breast, thyroid and lymph nodes. [3]. The appearance of metastases can only occur eight to sixteen years after diagnosis. [4].

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The radiological appearance of TFS is nonspecific. The tumor is hypervascular with a well-developed vascular network. [5]. We report a case of recurrence of meningeal TFS with pancreatic metastases. SFT of the pancreas is extremely rare, with around twenty cases reported in the literature [6.7].

Observation

A 57-year-old woman with a history of meningeal hemangiopericytoma that had undergone resection 18 years earlier presented for a brain MRI with symptoms of headache, which revealed a multifocal recurrence.

These were six extra-axial tumor formations, with a broad base of meningeal implantation, involving the left occipital operating site, coming into contact with the superior sagittal sinus and the right sinus, as well as the left frontal and right parietal lobes, presenting a intense heterogeneous intra-tumor enhancement. (Fig. 1).

A thoraco-abdomino-pelvic tomography scan as part of the extension assessment was carried out, revealing a total situs inversus and the presence of two masses involving the body and tail of the pancreas, well circumscribed with lobulated contours, measuring respectively 4, 1 x 4.5 cm and 4.2 x 4.5 cm, iso dense in spontaneous contrast without obvious calcification, and heterogeneous peripheral enhancement in the arterial phase, with progression in the portal phase, delimiting a weakly enhanced central zone suggestive of necrosis (Fig. 2).

There was no significant pancreatic ductal dilatation. No signs of engulfment of the adjacent vasculature were noted and no other foci of metastatic disease were observed in the abdomen.

Complementary magnetic resonance imaging (MRI) showed low signal of peripheral solid components on T1-weighted imaging and slightly higher signal intensity on T2-weighted imaging compared to the pancreatic parenchyma. The non-enhanced central contingent on the CT scan presented as an intense high signal on the T2-weighted sequence, confirming a cystic or necrotic component. On the diffusion sequences, the peripheral component appears in hypersignal b800, with clear restriction of diffusion and a low ADC coefficient. (Fig.3) In view of this morphological and dynamic aspect, we mentioned secondary locations of TFS or neuroendocrine tumors secondly.

A biopsy of the mass was performed and histopathology revealed a very dense cellular proliferation made of round or ovoid cells with no particular arrangement, large hyper chromatic nuclei without mitoses with a small stroma represented by vessels with clean walls sometimes dilated, compatible with a metastatic solitary fibrous tumor (high-grade hemangiopericytoma).

Immunohistochemical staining confirmed this diagnosis with intense positivity for CD34, CD99, and spot positivity for cytokeratin AE1/AE3.

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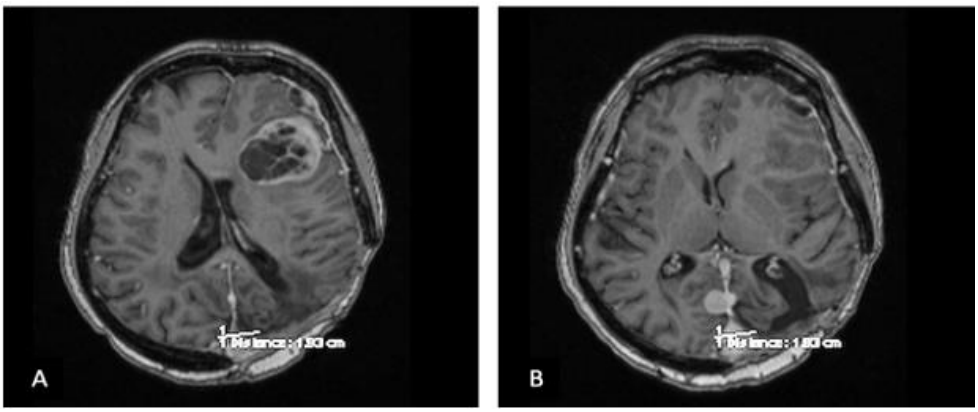

Figure 1: Magnetic resonance imaging (MRI) images of the described case demonstrating multifocal recurrence of intracranial meningeal SFT (A, B)

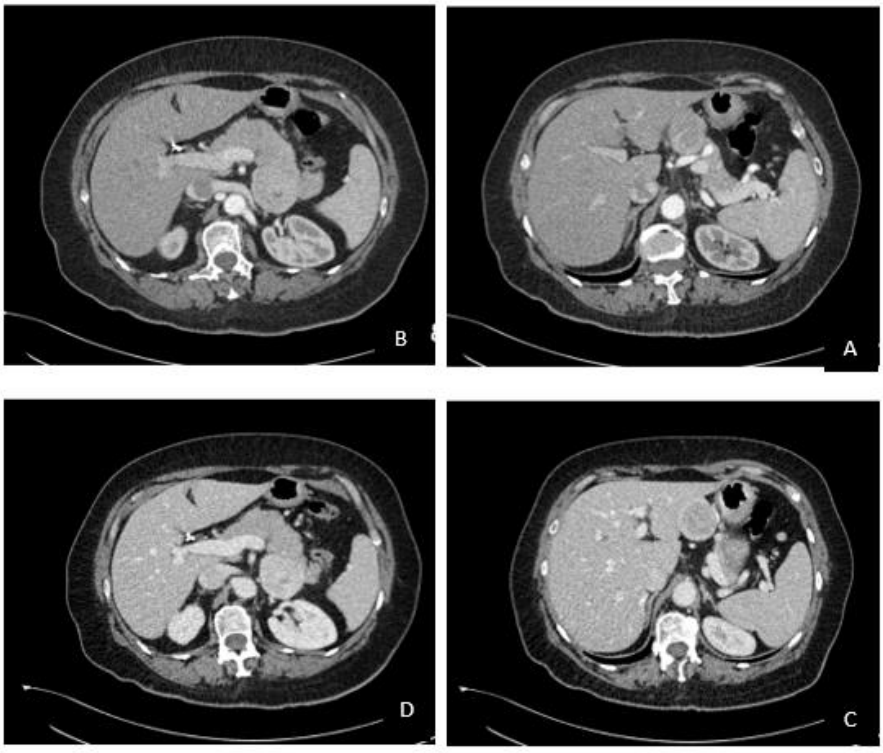

Figure 2: Dynamic contrast-enhanced computed tomography (CT) images of the described case demonstrating two solitary fibrous tumor metastases involving the body and tail of the pancreas. During the arterial phase (A, B), the mass appears well limited and is enhanced at the periphery with a non-enhanced central area. Subsequent venous phase imaging (C, D) shows progression of enhancement of the tumor edges, the masses are clearly demarcated from the pancreatic parenchyma and the central non-enhanced area is compatible with necrosis. The liver parenchyma adjacent to the body mass is compressed by the resulting mass effect.

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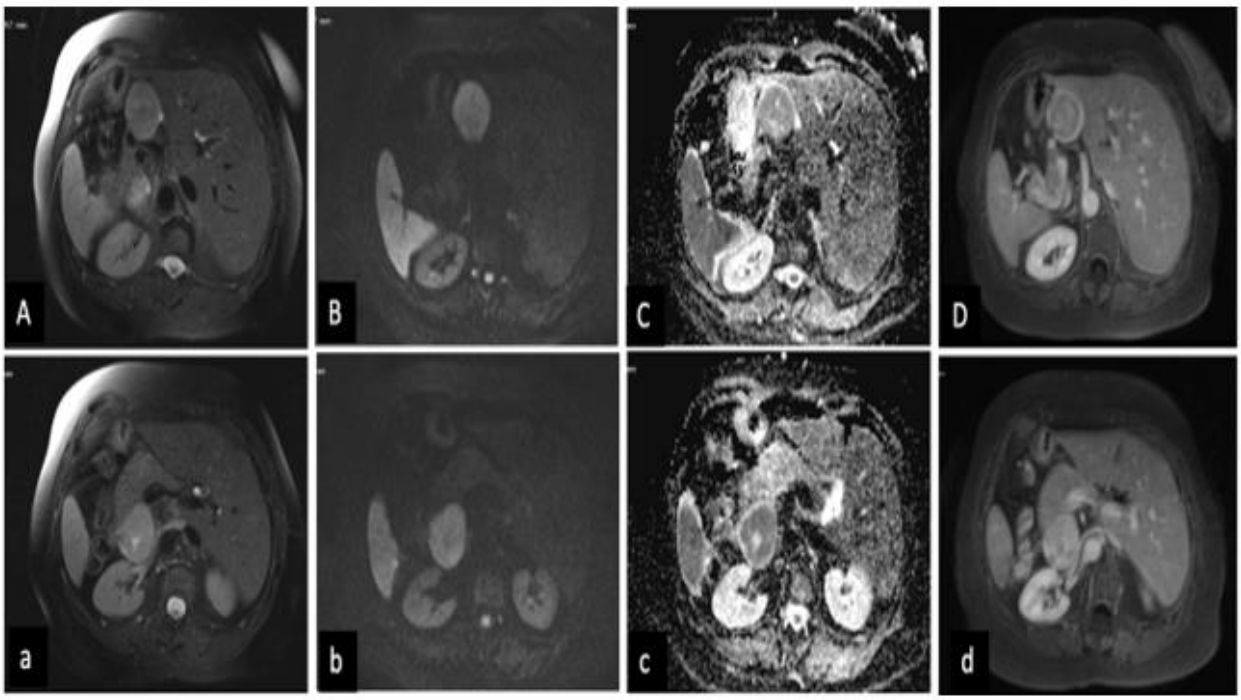

Figure 3: Images from magnetic resonance imaging (MRI). The peripheral solid component was hyperintense to the pancreatic parenchyma on T2-weighted imaging, and the central unenhanced portion was bright frank high signal indicating a cystic or necrotic component (A, a, D, d). Diffusion sequences showed a solid diffusion hypersignal component with decreased ADC (B, b, D, d).

DISCUSSION

TFS is a rare tumor arising from mesenchymal cells with a branched hemangiopericytoma-like vascular pattern, and was first reported in 1931 by Klemperer and Rabin; which mainly affects soft tissues and more rarely the meninges [5, 9]. TFS can develop in any organ containing mesenchymal tissue. It is generally observed in the thoracic cavity, but also extrathoracic locations such as in the central nervous system, orbit, sinus tract, thyroid gland, liver and kidney have been reported [6, 10- 15].

Intracranial HMPs clinically and biologically demonstrate aggressive behavior even after radical surgery and adjuvant radiotherapy. Local recurrence rates up to 20 years after primary resection of up to 90% and metastatic rates of up to 33% have been reported, hence the importance of close follow-up with imaging. [5,8,16].

The pancreas is an unusual location for this tumor, with 24 cases of metastatic pancreatic involvement after primary intracranial HMP in the English literature. [5,6,8]. In only 3 cases, the pancreas was the first metastatic site, and concomitant local recurrences were reported in 2 of 3 cases [5,6, 8, 17,18].

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The tumor is more common in women and usually occurs in the fourth or fifth decade [5, 6, 20].

The main symptom was pain, or jaundice, while 50% of patients were asymptomatic and incidentally diagnosed. The distribution of TFS localization in the pancreas is almost equal and nonspecific. [6] The imaging results of TFS are similar whether the tumor is intra- or extra-cranial. Alpern et al. Described a small series of abdominal TFSs and described features of a lobulated mass with enhancing solid components, cystic areas, and speckled calcifications as suggestive, but not specific, of a TFS. Finding of a hypervascular mass seems common in all reports. TFS have been described as well circumscribed or encapsulated without invasion of the parenchyma of the adjacent organ [1-19]. These characteristics were observed in the case described here.

The main differential diagnosis is neuroendocrine tumor. The differentiation between these lesions is mainly based on the clinical history. [5]

The MRI examination in general demonstrates a well-circumscribed, non-infiltrative mass, hypo- or isointense on T1-weighted imaging and hyperintense on T2-weighted imaging, as seen in the present case. In addition, without injection we can sometimes find fluid portions, which for the most part derive from cystic degeneration and rarely from tumor necrosis or retention cyst in frank hyper signal on the T2 sequence. However, these results are not specific characteristics of TFS [6].

TFS presents no specific clinical or imaging features, making preoperative diagnosis difficult. Generally, an accurate diagnosis can be established via pathological examination of specimens obtained by pancreatic biopsy guided by CT or endoscopic ultrasound imaging.

Histopathologically, TFS is composed of spindle cells developing in a hemangiopericytomatous system and exhibiting stroma with variations in collagen proportions [21]. Areas of necrosis, cystic or myxoid change, calcification, hemorrhage, increased vascularity, atypia, or malignancy may also be seen. Blood vessels often contain large-caliber, thin-walled branchial blood vessels [22].

However, histology sometimes cannot differentiate TFS from other spindle cell tumors. Immunohistochemical analysis remains essential to establish a definitive diagnosis of TFS. TFS typically stains positive for CD34, Bcl-2, and CD99, while it is typically negative for cytokeratin, epithelial membrane antigen, smooth muscle actin, desmin, S-100, c-kit, chromogranin, and synaptophysin. CD34, Bcl-2 and CD99 have been recognized as a representative marker of TFS, but they can also be positive in other neoplasms [6].

In this case, the tumor was positive for CD34 and CD99, which was a typical TFS finding.

Hemangiopericytoma (HMP), which was previously classified as a separate tumor entity, is now considered identical to TFS based on its immunostaining profile, and molecular and genetics. In the 2016 revised 4th edition of the WHO classification of CNS tumors,

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they were grouped under a single diagnosis (solitary fibrous tumor/hemangiopericytoma), and in the 2021 5th edition, the term hemangiopericytoma was dropped entirely [2].

CONCLUSION

The diagnosis of TFS on CT or MRI imaging, whether a primary soft tissue lesion or a metastasis from an intracranial primary, remains difficult despite multiple case reports of this entity. The tumor is rare and imaging findings are nonspecific and are similar to primary and secondary hypervascular tumors.

TFS should be considered as possibly representing metastatic disease in the face of an atypical hypervascular encapsulated pancreatic mass, particularly in patients with a history of intrathoracic or intracranial TFS.

Currently, a definitive diagnosis of TFS is based on immunohistochemical analysis on surgical specimens, but recent advances in immunohistochemistry make it possible to establish a preoperative diagnosis on biopsy samples.

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