A presacral solitary fibrous tumor with extramedullary hematopoiesis: radiologic and pathologic findings

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Abstract

Solitary fibrous tumors (SFT) are rare, ubiquitous neoplasms of mesenchymal origin, with distinctive histopathological and immunohistochemical features. We herein report an unusual case of a presacral SFT diagnosed in an asymptomatic 40-year-old woman preoperatively investigated with computed tomography and magnetic resonance imaging. Post-operative pathology examination showed a SFT containing foci of extramedullary hematopoiesis. Revision of preoperative imaging did not evidenced any findings suggesting this unusual association. The patient was free from local recurrence and metastases one year after operation. Differential radiological and histological diagnoses of solid presacral masses is briefly discussed.

Introduction

Solitary fibrous tumor (SFT) is a rare, ubiquitous lesion of a probable fibroblastic or myofibroblastic origin with a hemangiopericytoma-like vascular pattern, previously thought to arise from either mesothelial cells or submesothelial fibroblasts and hence restricted to mesothelium-lined surfaces. This tumor has been successively documented in almost every anatomic site, so presently pleural and extrapleural SFTs are now considered a single entity since they share similar pathologic morphology and immunophenotype.[1-3] Typical histopathological and immunohistochemical characteristics allow a confident preoperative diagnosis.[1,2,4]

We herein describe a case of a presacral SFT containing extramedullary hematopoietic foci an unusual feature never described in extrapleural SFT to the best of our knowledge.

Case Report

An asymptomatic 40-year-old woman was referred to our Hospital to investigate a pelvic mass detected by a gynecological ultrasonography in a private diagnostic center. She suffered from epilepsy well controlled with carbamazepine. Routine blood chemistry revealed a sideropenic anemia (RBC 3.60×10^{12}/L; Hb 9.7 μg/dL; MCV 73.9 fL; HCT 26.6%; MCH 26.9 pg; MCHC 35.1 g/dL; plasma-iron concentration 40 μg/dL; plasma-transferrin concentration 179 mg/dL), decrease of blood levels of sodium (129 mmol/L) and chlorine (90 mmol/L). Urinalysis was normal. Serum level of CA 15-3 was slightly elevated (43 U/mL; upper normal limit 32 U/mL); serum CA 125, CEA, Alpha-FP and CA 19-9 were within normal limits.

Computed tomography (CT) detected a presacral well demarcated lobulated mass, measuring 12.5×9×10 cm; the lesion was homogenously hypoattenuating relative to muscles on non-contrast enhanced scans and inhomogeneously hyperattenuating on contrast-enhanced scans; neither intratumoral fat nor calcium deposits were evident (Figure 1A). The mass displaced the rectum on the right side, the vagina and the uterus anteriorly, without evidence of local spread or erosion of the sacrum and the coccyx (Figure 1B).

The patient was then investigated with a 3T magnetic resonance imaging (MRI). The mass was homogenously isointense to the muscles on T1-weighted MRI (Figure 2A). On T2-weighted MRI the mass was well demarcated by a hypointense capsule and heterogeneous but hyperintense on T2-wheighted fat saturation images (Figure 2B); diffusion weighted images demonstrated a water diffusion restriction (Figure 2C). The mass strongly enhanced after intravenous gadolinium contrast medium administration; small non-enhanced foci and hypertrophic vessels were also appreciable (Figure 2D).

No nuclear medicine investigation was performed.

The patient was then operated on and rapidly recovered after surgery. She was free from local recurrence and metastases on the basis of a MRI and an whole body CT one year after the operation.

Grossly the tumor was well circumscribed, firm, homogeneous and brownish in color, measuring 10 cm in diameter on cut surface.

Microscopically it was a mesenchymal neoplasm characterized by ovoid or spindle cells with focal nuclear pleomorphism and by a prominent branching, *hemangiopericytoma-like* vascular pattern (Figure 3A). There was no evidence of necrosis and the mitotic activity was low (1 mitosis/10 HPF).

The immunohistochemical examination showed staining for CD34, bcl2 and vimentin, and negativity for desmin, smooth muscle actins, various cytokeratins, S100 protein, Synaptophysin and Chromogranins.

In addition the presence of sparse multinucleated cells prompted the search for extramedullary hematopoiesis, which was confirmed by specific immunostainings for megakaryocytes (CD61), precursors of granulocytes (myeloperoxydase) and of red blood cells (CD71) (Figure 3B).

Discussion

SFTs are mesenchymal neoplasms mainly found in adults with a mean age in the fifth decade, without a gender predilection.[1-4] Many SFTs were previously classified as hemangiopericytomas or pericytoma-like tumors but, because cells of hemangiopericytomas show a fibroblastic differentiation rather than a pericytic feature, the term hemangiopericytoma was progressively abandoned. It is now accepted that *haemangiopericytoma* simply represents a heterogeneous group of different neo-

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plasms (including SFTs) sharing a common growth pattern, and this term should be abandoned.

Morphologically SFTs show a spectrum of appearances, which include a fibrous and a cellular variant, as well as fat-forming cases , a giant-cell rich variant and some overlapping cases with deep fibrous histiocytoma.[3] Many tumors are found along serosal surfaces, in the head and neck, extremities, abdominal wall, back, buttock and perineum, groin, vulva and retroperitoneum.[1,2,5-7]

SFTs are slow-growing and incidentally detected in up to 50% of cases.[2,3] Symptoms are related to a mass effect and depend on the anatomical localization;[2,3] paraneoplastic hypoglycemia (Doege-Potter syndrome) is found in 5% of patients.[1,2,4,5] The behavior of extrapleural SFTs is unpredictable. Roughly 10-15% of these tumors show malignant behavior in the form of recurrence or metastases. Criteria for malignancy include size (greater than 5 or 10 cm according to the authors), necrosis, nuclear pleomorphism and mitotic activity index (>4 mitoses per10 HPF). However the relationship between morphology and outcome is poor in SFT and some (rare) morphologically innocuous lesions behave aggressively.[1,2]

The case herein described had a large size and focal nuclear pleomorphism, but no other morphologic criteria of malignancy, and in particular no evidence of necrosis and a low mitotic activity.

A peculiar feature of this tumor is the presence of extramedullary hematopoiesis, which was previously described in a single case of a SFT of the pleura, but never in extrapleural sites at the best of our knowledge.[8]

Complete surgical excision is curative but the tumor can recur after surgery; distant metastases usually involve lung, liver and bone.[3]

All SFTs share similar features on diagnostic imaging regardless of the origin site.[7,9,10] Presacral SFTs are usually large at the time of detection and exert a mass effect on neighbor organs and structures; they appear well-circumscribed, with smooth or lobulated contour, and discrete margins on CT and MR images. SFTs have an intermediate signal on both T1-weighted MRI and T2-weighted MRI; in the latter sequence, however, signal intensity depends on cellularity, fibrosis, cystic changes, necrosis.[7,9-11] SFTs strongly enhance after intravenous contrast injection and are frequently associated with hypertrophic vessels; hypoenhancing or nonenhancing foci are related with necrosis or cystic changes.[7,10,11] Calcium deposits may be found in large or malignant tumors.[7,9-11] A fatty component is recognizable in the fat-forming variant of SFT.[12] Diagnostic imaging can also demonstrate local invasion and distant metastases.[7]

Noninvasive differential diagnosis of a presacral mass include many non-cystic lesions enhancing on CT scan or MRI. Although histology is mandatory for the diagnosis, narrowing of a preoperative differentiation can be clinically relevant in some instances, since role of percutaneous biopsy is debated and usually restricted to patients candidate to treatments other than surgery.[12,13]

Primary retrorectal malignant neoplasms (adenocarcinoma or sarcomas) and metastases appear isointense to muscles on T1-wheighted MRI, hyperintense on T2-wheighted MRI and enhance after contrast medium intravenous injection. Malignant lesions may have irregular borders and can cause bone erosion for local spread.[7,14,15] Presacral carcinoid usually arise from the rectal wall or a dysembryogenetic cyst.[12,16]

Retrorectal gastrointestinal stromal tumors (GIST) can rise from the rectal wall but they exceptionally found out from the digestive tract; in contrast with SFTs they show a high signal intensity, sometimes heterogeneous, on T2-wheighted MRI.[14,15]

Ovarian Brunner tumor and fibrothecoma

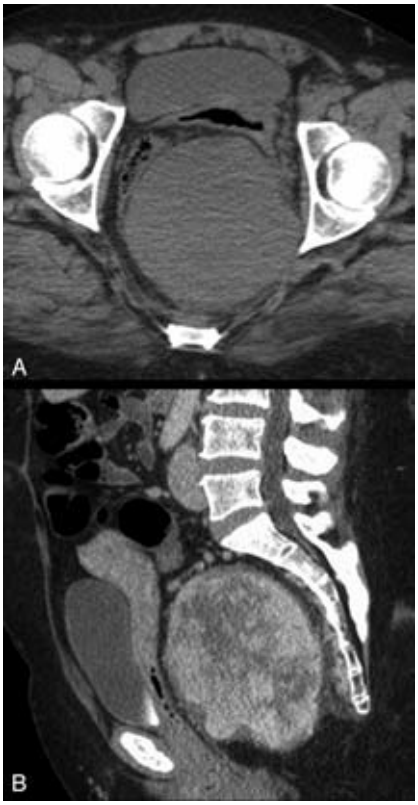

Figure 1. A) Non-enhanced CT shows a huge presacral homogenous mass displacing the vagina anteriorly and the rectum on the right side. B) Sagittal multiplanar reformation of contrast-enhanced CT clearly depicts a fat plan between the heterogeneously enhancing mass and the sacrum.

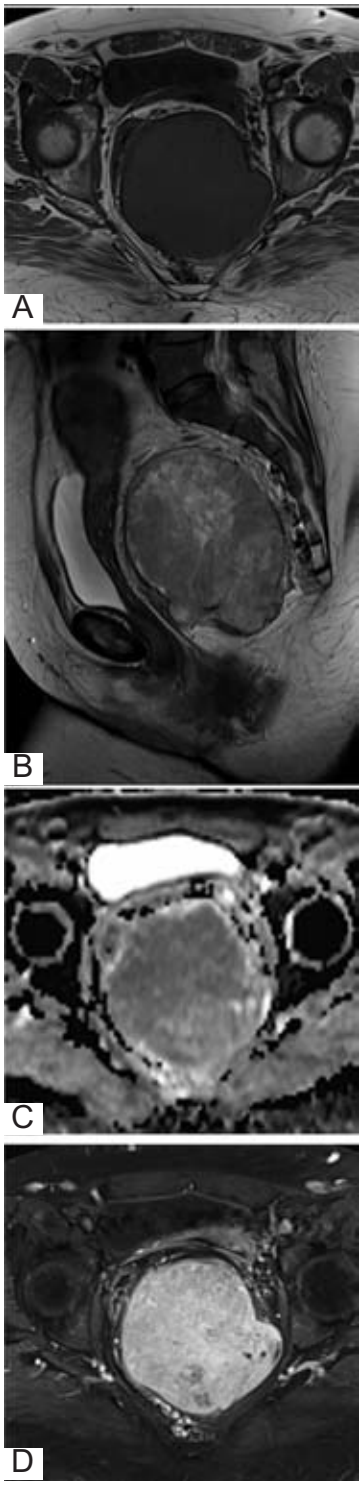

Figure 2. A) The mass present intermediate signal on T1-weighted MRI; tiny linear structures of voiding signal, due to hypertrophic vessels, are recognizable within the lesion. B) The lesion is heterogeneous on sagittal T2-weighted MRI. C) The mean apparent diffusion coefficient value measured on ADC map is 1210×10^{-3} mm²/s. D) The mass strongly enhances after intravenous gadolinium injection. Spots of signal void and small non-enhancing areas, corresponding to high flow vessels and fibrotic changes respectively, are also appreciable.

exhibit a low signal intensity on T2-wheigetd MRI and a delayed enhancement on contrast-enhanced imaging; they usually hide the ovary which they originate from.[9] A similar MRI pattern is found in pedunculated uterine leiomyomas or primary retrorectal leiomyomas rising from müllerian or wolffian remnants.[9,15]

Flow void signal and a vascular enhancement on MRI are typical of congenital pelvic arterio-venous malformations. Mixomas, schwannoma and plexiform neurofibromas are hyperintense on T2-wheighted MRI.[15] Plexiform neurofibromas typically exhibits a target-like pattern on T2-wheighted MRI and on post-contrast images, a feature sometimes detectable in schwannomas. Intratumoral mixoid changes of fatty accumulation appear hypoattenuating on CT scans and non-enhancing after intravenous contrast injection.[15]

Paracordomas share CT and MRI features similar to those of SFTs but can invade the sacrum; association with hypertrophic vessels is not reported.[13,17]

Presacral fat-forming variant of SFTs must be differentiated from extramedullary hematopoiesis, liposarcoma, teratoma and myelolipoma.[15] Presacral extramedullary hematopoiesis are characterized by a fatty components and an intermediate signal intensity on T2-wheighted MRI, which make these lesions radiologically indistinguishable from the fat-forming variant of SFTs; nevertheless a diagnosis of an extramedullary hematopoiesis is relevant because surgery is indicated only in symptomatic patients. A presacral extramedullary hematopoiesis should be considered in patients with bone marrow disorders or hemolytic anemia; furthermore extramedullary hematopoiesis is hyperactive on bone marrow ^{99m}Tc sulfur colloid scintigraphy.[18-20]

Liposarcomas usually exhibits infiltrative growth. Myelolipoma is well circumscribed and more frequently found in elderly women, but it is indistinguishable from a SFT on the basis of the imaging.[15] Teratoma is commonly detected in younger patients and show a complex pattern on imaging depending on tissue arrangement.[12,15]

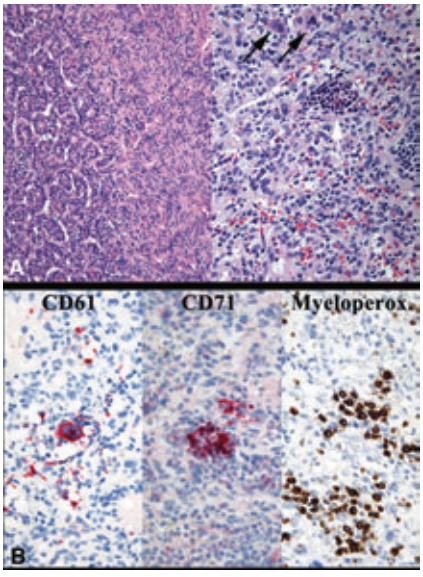

Figure 3. A) Left panel (H&E, 20×): typical vascular pattern of the SFT. Right panel (H&E, 40×): SFT with extramedullary hematopoiesis (black arrows: megakariocytes; white arrow: an erythroid island). B) Extramedullary hematopoiesis: immunohistochemical staining highlighting megakariocytes (CD61), nucleated erythroid cells (CD71) and granulocyte precursors (myeloperoxydase).

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