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LETTER TO THE EDITOR

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Epithelioid solitary fibrous tumor in the ischioanal fossa

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Dear Editor:
Solitary fibrous tumor (SFT) is a rare mesenchymal tumour initially described in the pleural cavity, but later reports have documented its occurrence in various extrapleural sites, including the abdomen [3, 5, 6]. The diagnosis of these tumors has generally been fraught with difficulties, because of their histologic variability. They can display storiform, hemangiopericytic, herring-bone, neural-type palisading, and diffuse sclerosing areas [4].

We describe the second case of SFT in the ischioanal fossa, characterized by epithelioid areas. To our knowledge, this feature has not been documented in this location before.

A 67-year-old male, with unremarkable medical history, presented with urinary problems. Rectal examination revealed a fullness in the right side of the rectum. MRI study showed the presence of a 14×8 cm moderately circumscribed, heterogeneous solid mass, with high vascularity, and central necrosis, occurring in the right ischioanal fossa (Fig. 1). This mass was extra-peritoneal in location, with resultant superio-left displacement of the levator musculature and the rectum. The tumor was totally excised. No further therapy was given, and the patient recovered well, without evidence of recurrence as yet (6 months of follow-up).

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Grossly, the excised lesion was a well circumscribed, partially encapsulated, rounded mass, measuring 13×12×8 cm. On section, the mass had multinodular, gray-whitish and firm appearance, with myxoid and hemorrhagic changes at the center (Fig. 2).

Microscopically, the tumor was characterized by prominent vascularity, with numerous small and mid-sized caliber vessels, with hyalinized wall "hemangiopericytoma-like" and significant variation in cellularity (Fig. 3). Hypercellular areas were predominant at the periphery, they displayed compact clusters of epithelioid neoplastic cells, with abundant eosinophilic cytoplasm, and distinct cell borders (Fig. 4). The nuclei were round with finely dispersed chromatin and small nucleoli. There was no nuclear atypia and mitoses were difficult to find (<1/10 HPF). Necrosis was prominent at the center, as well as myxoid degeneration. There was no storiform pattern, neuroid appearance, or diffuse sclerosing areas, even after multiple sampling of the tumor. The surgical margins were found to be clear.

Immunohistochemically, tumor cells were strongly positive for vimentin, CD34 (Fig. 3, inset), BCL2 (Fig. 4, inset), and moderately for CD99 (MIC 2). Scattered cells reacted with KL1. Immunostains for EMA, actin, desmin, CD117 (c-Kit), and S100 protein, were negative.

The diagnosis of epithelioid SFT, with unpredictable prognosis was made. SFT is an uncommon spindle cell neoplasm that was first described as a distinctive pleural lesion in 1931. In subsequent decades, extra-thoracic SFTs have increasingly been recognized. In this paper, we report the second case of SFT arising in the ischioanal fossa; the first was described by Dudkiewicz et al. in 2004 [2]. What makes the present lesion apparently unique, is the presence of epithelioid features. This morphologic variability may contribute to more difficulties in diagnosing these tumours, especially on percutaneous biopsy.

Histologically, these tumours are characterized by a variety of growth patterns, with admixture in a various proportion of bland spindle cell cytology, alternating hyper- and hypocellular areas, keloid-like hyalinisation, and a frequently prominent branching vasculature often described as

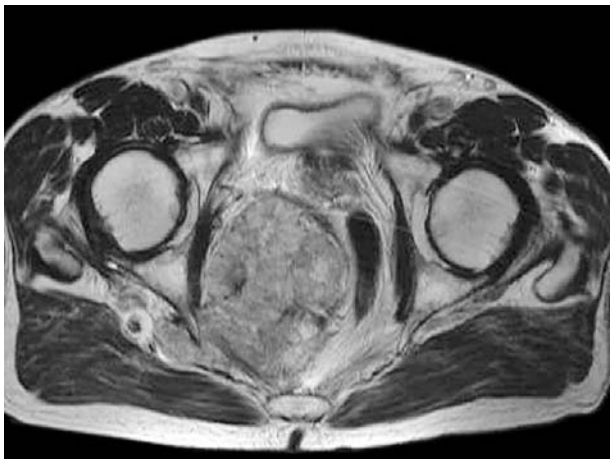

Fig. 1 RMI : 14 cm moderately circumscribed, heteregeneous solid mass, with high vascularity, and central necrosis

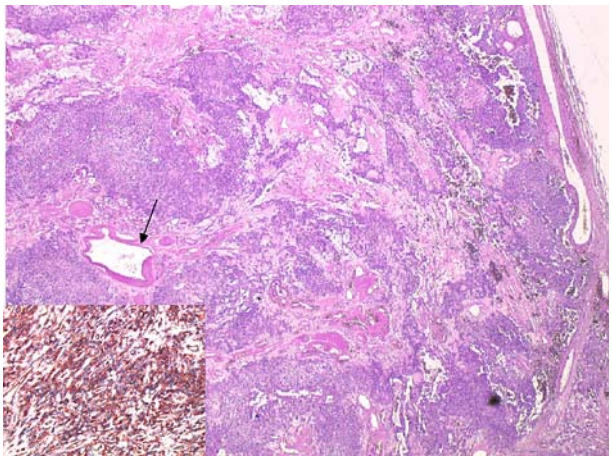

Fig. 3 Well circumscribed tumor, with numerous small and mid-sized vessels, with hyalinized wall (arrow) "hemangiopericytoma-like" and variation in cellularity. Strong immunostaining positivity of the tumor cells with CD34 (inset)

"hemangiopericytoma-like". Areas showing an angiofibroma-like appearence, neural-type fascicle areas, with wavy nuclei, and occasional herring-bone patterns are also reported [5]. Other less frequently observed cytological features are nuclear palisading, compact clusters of polygonal cells, and small cells [4].

All these patterns were absent in our observation, which was composed entirely of epithelioid cells. The term "epithelioid SFT" was proposed for the first time in 2003 by Alberto et al., when they reported a mediastinal SFT, with predominantly epithelioid cells, exhibiting histopathological and immunohistochemical features intermediate between those of SFT and those of cellular adenomatoid tumor [1]. Unlike this observation, SFT was entirely composed of epithelioid cells in our case, and only scattered tumor cells were positive for cytokeratin.

This epithelioid growth pattern contributes to further difficulties in differential diagnosis of these tumors with other soft tissue neoplasms, with epithelioid features, such as epithelioid sarcoma, schwannoma, leiomyosarcoma, and malignant gastrointestinal stromal tumor. Immunohistochemical stains will be of great value by demonstrating positivity of epithelioid cells with vimentin, CD34, BCL2, and CD99, and their negativity with CD117 (c-Kit), epithelial, neural, and muscular markers.

The behaviour of extrathoracic SFTs is unpredictable, comparable to their pleural counterparts, although they have an increased risk of local recurrence. Positive surgical margins, tumor size greater than 10 cm, and the presence of malignant component (necrosis, marked cellularity, nuclear atypia, and more than four mitoses/10 HPF) predict worse-metastasis-free survival [3]. Whether this epithelioid pattern represents another atypical histological features SFT awaits further studies with larger numbers of cases.

In summary, we report a case of epithelioid SFT arising in the ischioanal fossa, which highlights the difficulties encountered in the diagnosis of SFTs because of their histologic variability. One should bear in mind that epithelioid features may exist, and could be prominent in some

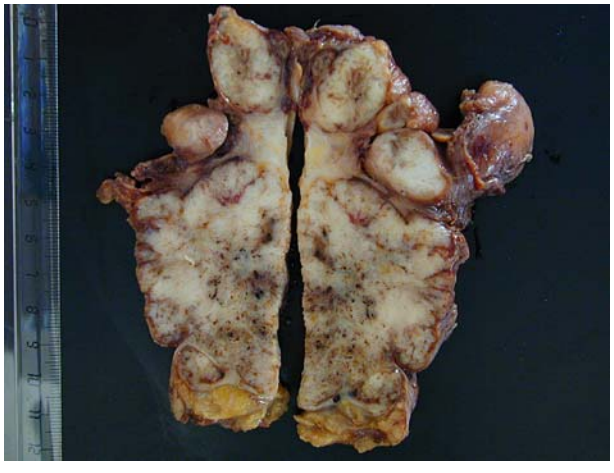

Fig. 2 Gross section : multinodular, gray-whitish mass, with myxoid and hemorrhagic changes at the center

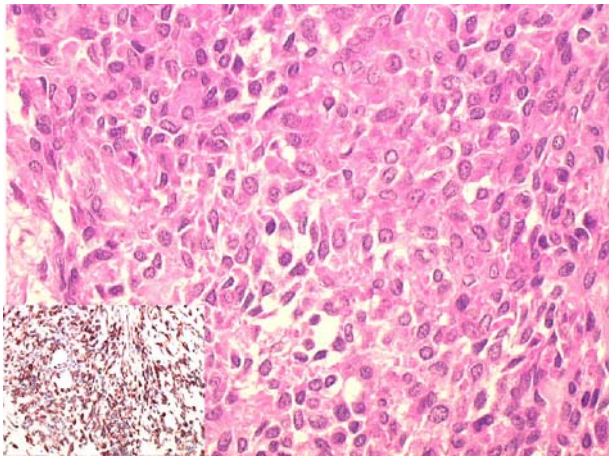

Fig. 4 High power view of epithelioid tumor cells, with abundant eosinophilic cytoplasm, distinct cell borders, and round nuclei, without atypia and mitosis. Diffuse immunostaining positivity of tumor cells with BCL2 (inset)

cases. Our observation emphasizes the importance of an appropriate immunohistochemical panel in the differential diagnosis of epithelioid cell neoplasms in the pelvis. Identification of this pattern of SFT is of importance, to avoid misdiagnosis with other more aggressive conditions in the soft tissue location.

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