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Case Report

Angioleiomyoma of the lumbo-sacral region with unusual keloid-like collagen fibers

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Summary
Angioleiomyoma is a benign soft tissue tumor which usually occurs in superficial or deep soft tissues. Only rarely does this tumor occur at unusual sites, including retroperitoneum. We present a rare case of lumbo-sacral angioleiomyoma in a 54-year-old man. Apart from this unusual site, the most striking morphological feature was the presence of numerous keloid-like collagen fibers interspersed among the fascicles of the neoplastic cells. Radiological, morphological and immunohistochemical features are presented, and differential diagnosis with its potential morphological mimickers is discussed.

Key words: angioleiomyoma, retroperitoneum, lumbo-sacral region, keloid-like collagen fibers

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Conflict of interest
The Authors declare no conflict of interest.

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Introduction

Angioleiomyoma (ALM), also known as "*vascular leiomyoma*", is basically a leiomyoma which arises from the smooth muscle cells of intratumoral thick-walled vessels. Although this tumor usually occurs in the subcutaneous tissue, especially of the lower extremities, other unusual sites, including the head and neck region, uterus, ovary, and the broad ligaments, can be involved [1-6]. A PubMedline-based search of the English language literature revealed that ALM of the retroperitoneum are exceptionally rare, with only 4 cases reported so far [7-10]. We herein report a rare case of ALM occurring primarily in the lumbo-sacral region of a 54-year-old man. Apart from the unusual site, the most striking morphological feature was the presence of numerous keloid-like collagen fibers interspersed throughout the tumor.

Clinical findings

A 54-year-old man was complaining of a vague lumbo-sacral pain for about three months. CT scan and MR showed a solid mass measuring 7 cm in its greatest diameter, located in the deep soft tissues of the lumbo-sacral region (Fig. 1 A, B). A radical surgical excision of the mass was performed. Grossly, a nodular tumor mass with well circumscribed borders was seen. The cut surface showed a tan-gray multinodular mass with pseudocystic hemorrhagic areas (Fig. 1C). Histologi-

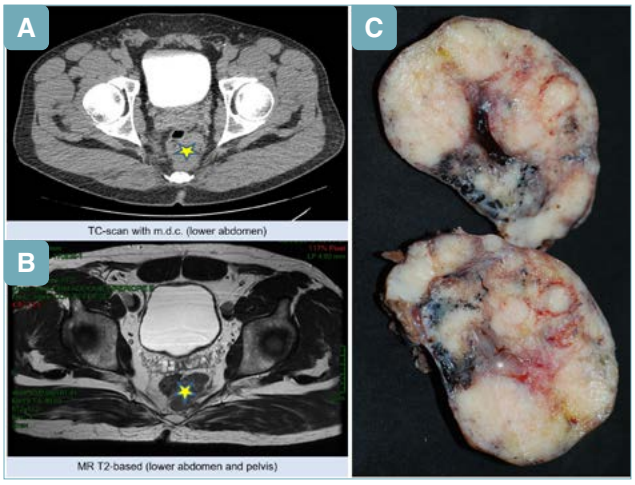

Figure 1. Radiological features: TC (A) and MR (B) showing a solid lumbo-sacral mass (yellow stars). (C) The cut surface of the mass showed a multinodular appearance with pseudocystic hemorrhagic areas.

cally, the tumor was mainly composed of interlacing fascicles of spindle cells with numerous interspersed keloid-like collagen fibers (Fig. 2). These areas were closely reminiscent of a mammary-type myofibroblastoma. The spindle cells had a bland-looking appearance, with eosinophilic cytoplasm and oval-shaped nuclei without nucleoli. Hypercellular areas were alternating with hypocellular edematous areas in which neoplastic cells showed a stellate morphology with anastomosing cytoplasmic processes, with a close resemblance to the reticular variant of perineurioma

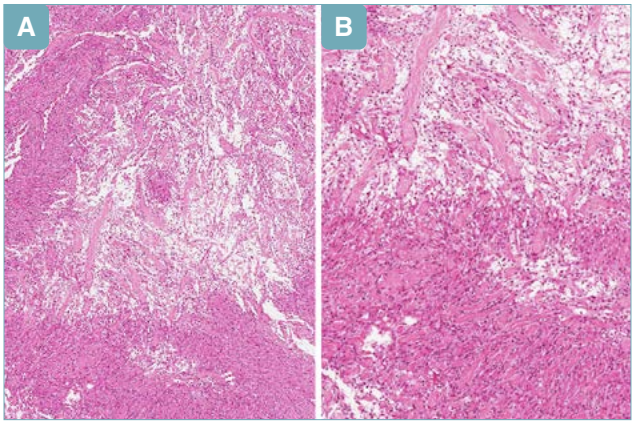

Figure 2. (A) Low-magnification showing alternating hypercellular and hypocellular areas, with interspersed keloid-like collagen fibers. (B) Higher magnification: spindle cell tumor with keloid-like collagen fibers.

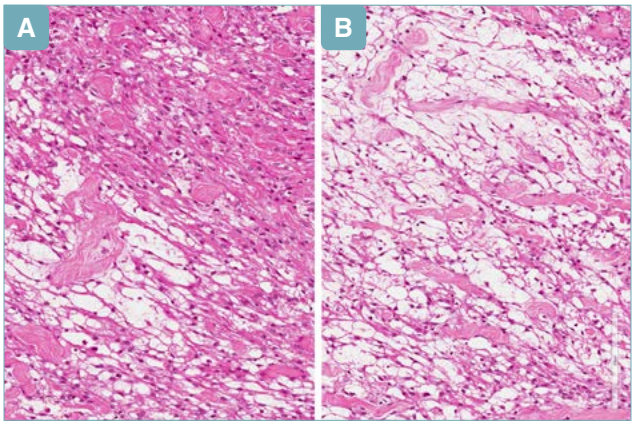

Figure 3. (A) Transition from hypercellular to hypocellular area. (B) The hypocellular areas were reminiscent of the reticular variant of perineurioma: the neoplastic cells adopted a stellate morphology with anastomosing cytoplasmic processes; notice the interspersed keloid-like collagen fibers.

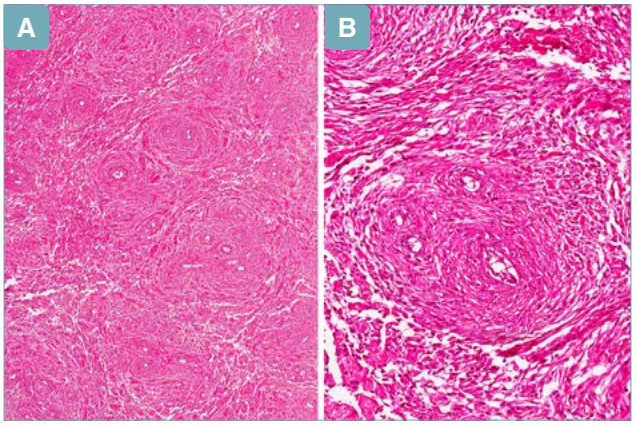

Figure 4. (A) Tumor area with the typical features of angioleiomyoma: a spindle cell tumor with fascicular growth pattern, exhibiting thick-walled blood vessels. (B) The smooth muscle cells of the vascular walls seem to blend into the spindle cells of the tumor.

(Fig. 3). In some areas (about 30% of entire tumor) numerous thick-walled venous-type vessels were seen (Fig. 4). At low magnification the neoplastic spindle cells exhibited a concentric, perivascular accentuation (Fig. 4A); however, at higher magnification, the smooth muscle cells of the outer layers of the vascular walls seemed to blend imperceptibly with the intervascular spindle cells (Fig. 4B). Nuclear atypia, mitoses and necrosis were absent. Immunohistochemically the spindle cells were strongly and diffusely stained with myogenic markers (desmin, α-smooth muscle actin

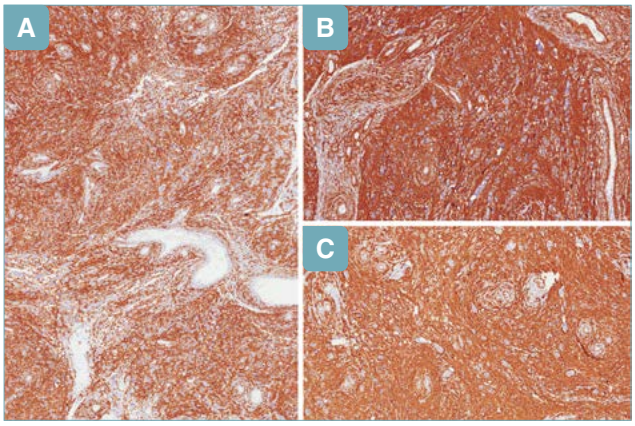

Figure 5. (A) Immunohistochemically, neoplastic cells are diffusely stained with desmin (A), α-smooth muscle actin (B) and h-caldesmon (C).

and h-caldesmon) (Fig. 5). No staining was obtained with CD34, CD117, DOG1, S100 protein, pancytokeratin, EMA, HMB45, STAT6, myogenin, CD99, or p63. Based on the morphological and immunohistochemical features the diagnosis of "*solid and venous-type angioleiomyoma with unusual keloid-like collagen fibers*" was rendered.

Discussion

ALM of the retroperitoneum is extremely rare with only a few cases reported in the literature [7-10]. Radiologic imaging is not specific, showing a solid nodular mass with well circumscribed borders. Accordingly, its diagnosis is histologically based, with immunohistochemical confirmation. Although the diagnosis of ALM is straightforward in appropriate sites (superficial/deep soft tissues), its unusual localization can pose serious diagnostic problems. Apart from the unexpected site (pre-sacral region) for an ALM, the most striking finding of the present case was the presence of numerous thick, keloid-like collagen fibers interspersed among the neoplastic cells. To the best of our knowledge this morphological feature has not been previously recognized/emphasized. As these keloid-like collagen fibers can be encountered in mammary-type myofibroblastoma, leiomyoma, solitary fibrous tumor and gastrointestinal stromal tumor (GIST), a differential diagnosis with these spindle cell tumors is mandatory. Apart from the breast parenchyma [11], mammary-type myofibroblastoma can occur in the soft tissues with a similar morphology [12]. Unlike ALM, it lacks the thick-walled vessels and the perivascular condensation of the spindle cells, while the neoplastic cells are usually CD34-positive and h-caldesmon-negative. Leiomyoma may show similar morphology, but its vessels are usually capillaries, along with a few arterioles and small arteries, in contrast to the thick-walled vessels of the ALM. Solitary fibrous tumor can occur at unusual sites [13-18] and rarely may present as a retroperitoneal/pelvic mass [19 20]. Unlike ALM, it shows a hemangiopericytomatous vasculature, with a typical branching configuration and perivascular hyalinization. In addition solitary fibrous tumor is stained with CD34 and STAT6, while it does not express myogenic markers. Only rarely do GIST occur as retroperitoneal masses [21] but, unlikely ALM, it stained with CD117, DOG1, and CD34.

In our case correct diagnosis was based on the identification, at least focally, of thick-walled blood vessels with smooth muscle cells which seem to blend imperceptibly with the intervascular smooth muscle cells of solid tumor areas. Immunohistochemistry, revealing a diffuse and strong staining for myogenic markers (α-smooth muscle actin; desmin; h-caldesmon), was helpful for confirming a diagnosis of ALM, ruling out potential morphological mimickers. The present case contributes to widen the morphological spectrum of ALM, emphasizing that this tumor may contain numerous thick, keloid-like collagen fibers interspersed among the neoplastic smooth muscle cells. The diagnosis of retroperitoneal ALM can be rendered confidentially if the pathologist is aware that this tumor can arise at this unusual site.

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