Leiomyoma of the uterus and retroperitoneal angioleiomyoma: case report

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Summary

Retroperitoneal angioleiomyoma is a benign mesenchymal neoplasm that is composed of smooth muscle cells and thick-walled vessels. In a 36-year-old patient a retrouterine and retroperitoneal tumor, 70 x 65 x 50 mm in size, was discovered during a surgical procedure due to uterine myoma. The tumor had a soft consistency and was completely removed. Histopathology showed features of angioleiomyoma. Angioleiomyoma is a rare benign entity; hence a benign course and good prognosis are expected.

Key words: Retroperitoneal angioleiomyoma; Uterine myoma.

Introduction

Angioleiomyoma is a rare benign tumor, occurring most frequently in adults and is composed of irregularly admixed mature smooth muscle fibers and thick-walled vessels. Angioleiomyoma has been described as a cystic tumor of the upper abdomen [1], more than as pulmonary lymphangiomyomata [2], and as lymphangiomyomatosis and angioleiomyoma of the uterus [3, 4]. Angioleiomyoma is usually found in the skin of the lower extremities though it is also known as a tumor appearing on the arms [5], urethrae [6], ear [7], retropharyngeal area [8] and kidneys [9]. The described retroperitoneal tumors were mostly incidental findings during other operative procedures and seemed to be quiescent. They were usually large tumors, at least 7 cm in diameter in most of the reported cases. We present case of a retroperitoneal angioleiomyoma (also discovered during a surgical procedure) in a woman with uterine myoma.

Case Report

A case of 36-year-old patient with uterine myoma which was situated on the front wall (about 65 mm in size) and pressing on the bladder so strongly that it brought about diuresis disorders. Although the menstrual cycle in our patient was normal she was advised to have an operation.

During the surgical procedure, which was performed by laparotomy, once the myomectomy had been performed and the uterus reconstructed, the pelvic area was examined. A tumor, 70 x 65 x 50 mm in size and of soft consistency (similar to bowel diverticulum), was found in the area of Douglas pouch. Being clearly limited, the tumor was completely removed and sent for pathohistological analysis. Postoperative recovery of the patient was normal and she was released from hospital five days following the surgery.

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Macroscopically, the tumor was well circumscribed with a thin fibrous capsule and the consistency was soft. The cut surface was yellowish-white in color. Microscopic examination (Figures 1 and 2) showed mixed cellular proliferation of mostly ovoid or slightly elongated smooth muscle cells showing no signs of epithelial cells. What was dominant in the histological image was a nodular and solid trabecular organization in a periadventitial zone of multiple, dilated, thick-walled blood vessels. Presence of much ectatic space conveyed an image of cavernous angiomatosis. Here and there, tumor areas conveyed an image of mixoid transformation. There *was* no necrosis. The mitotic index was 0/50 HPF and the Ki-67 index around 2%. Vascular and spindle cell components were immunoreactive for vimentin and smooth muscle acting with no reactivity to desmin and S100. The final histopathologic diagnosis was angioleiomyoma.

Discussion

Angiomyomas or vascular leiomyomas are benign soft tissue tumors composed of variable amounts of benign smooth muscle fibers, thick-walled vessels and stromal cells, namely adventitial fibroblastic cells. They usually develop between the age of 40 and 60. Typically, they cause few problems apart from pain. They may be small and painless when located in the head and neck region. [11]. However, pain is a dominant clinical feature if the tumor is situated in the abdominal region, especially if it is large in size. Apart from pain a dominant symptom is bleeding when angioleiomyoma develops in the uterine region, which is quite rare [4]. If the tumor is positioned in the retroperitoneal region it is soften quite inactive and is usually discovered during a surgical procedure performed for different reasons, as was the case with our patient.

The main differential diagnoses of angioleiomyoma include angiomyolipoma, leiomyoma with fatty degeneration, myolipoma, well-differentiated liposarcoma, spindle-cell lipoma, lipoleiomyosarcoma, and leiomyosarcoma.

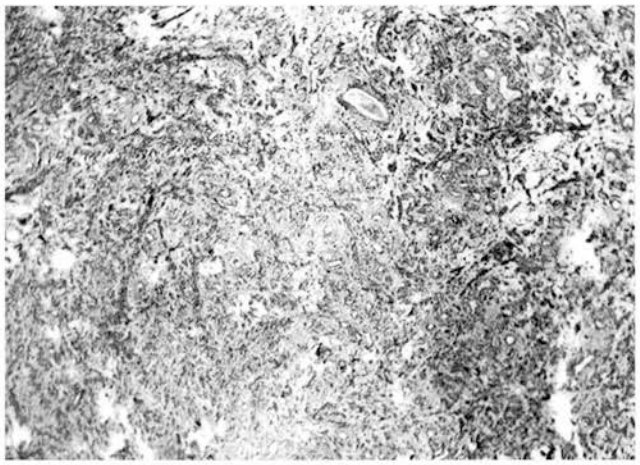

Fig. 1

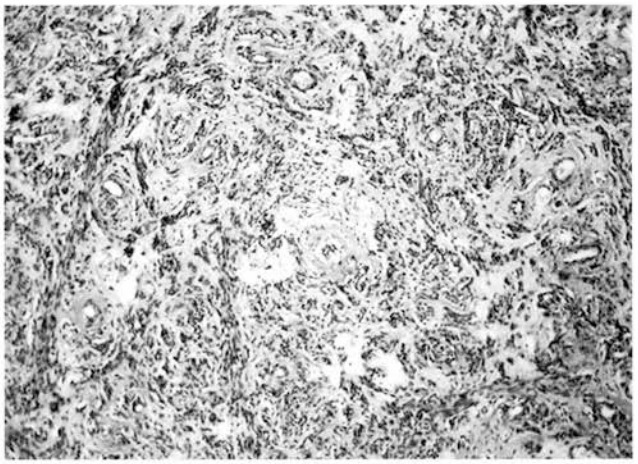

Fig.

Figure 1. — Nodules of smooth muscle fibers admixed with numerous thick-walled vessels (H&E x 40).
Figure 2. — Microscopic examination revealed numerous thick-walled vessels interspersed between smooth muscle cells (H&E x 40).

Angiomyolipomas contain conspicuous vessels showing thick muscular walls are HMB45-positive and frequently associated with tuberous sclerosis. A leiomyoma with fatty degeneration consists of both the components being heterogenously distributed within the mass with the adipose component focally distributed and not an integral part of the lesion. Tumors consisting of a mixture of mature adipose and smooth muscle tissues, including those designated lipoleiomyomas, fibrolipoma leiomyomas and myolipomas, are exceedingly rare. Most commonly the muscular component is predominant. Soft tissue myolipoma is a benign lesion, which has to be distinguished from lesions with malignant or uncertain biologic behavior. Leiomyosarcomas of the retroperitoneum invading adipose tissue contain an abundance of mitoses in the smooth muscle component and presence of recurrence or metastasis on follow-up.

The origin of angioleiomyoma is unclear. Angioleiomyomas are classified into three histological types: capillary or solid, cavernous and venous. Angiomyomas are believed to arise from the vessel walls (10). In the present study, fibroblastic cells were detected at adventitial areas of vessels. These results suggest that angiomyomas may originate in vessel walls, and that the thick-walled vessels within angiomyomas are different from normal vessels.

Conclusion

None of the angioleiomyomas reported have shown recurrence or metastasis. Neither did our case. Thus a benign course and good prognosis are expected. Although angioleiomyoma is very rare, pathologists should consider it in the differential diagnosis of muscle-containing retroperitoneal masses.

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