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Spermatic Cord Cellular Angiofibroma

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Synonyms

Angiomyofibroblastoma-like tumor

Definition

Rare, circumscribed, slow-growing, benign mesenchymal tumor whose suggested histogenesis is perivascular stem cells.

Clinical Features

- **Incidence**
  Very low. About 60 cases of cellular angiofibroma in male genitourinary system are reported in English literature.
- **Age**
  It affects adults usually over 50 years of age.
- **Site**
  In men, cellular angiofibroma is observed in the inguino-scrotal region (paratesticular, tunica vaginalis, epididymis, and spermatic cord). There are rare reports of perineal, urethral, and retroperitoneal locations.
- **Treatment**
  Recommended treatment is the complete surgical excision with tumor-free margins and long-term follow-up exams. No indications for radiation and chemotherapy.
- **Outcome**
  Follow-up is required because it may recur years after initial resection. No metastases have been observed.

Macroscopy

Macroscopic appearance is of a well-circumscribed, elastic-hard, slightly mobile mass whose size varies between 2.5 cm and 14 cm with an average of 6.7 cm. The cut surface is edematous and the parenchyma is cream color with clear edges and focal mixoid areas.

Microscopy

The tumor consists of two principal components (Fig. 1):

– Spindle-shaped cells with eosinophilic cytoplasm and no cytological atypia;
– Numerous irregular, round, small- to medium-sized, thick-walled blood vessels. They are often hyalinized.

H. V. Krieken (ed.), *Encyclopedia of Pathology*,
https://doi.org/10.1007/978-3-319-28845-1_4942-1

The surrounding stroma is edematous with variable amounts of collagen bundles. In the background, many mast cells and lymphoid cells forming clusters have been described. Some cases showed intralesional fat and myxoid change. Necrosis or atypical mitotic figures are absent. A fibrous pseudocapsule is not always visible.

Immunophenotype

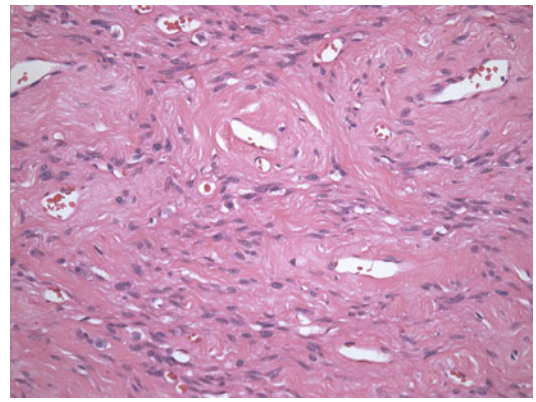
The spindle cells are usually positive for CD99 and vimentin. In more than half of the reported cases, the tumor cells are positive for CD34 and are variably positive with desmin and smooth muscle actin. They are negative to cytokeratin, protein S-100, EMA, myogenin, inhibin, C-kit (CD117), and calretinin. Stains for Factor VIII, CD31, and CD34 are also negative in the spindle-shaped cells but positive in the endothelial cells within the numerous irregular vessels.

Molecular Features

The loss of *RB1* and *FOXO1A1* genes due to the deletion of the 13q14 region in cellular angiofibroma is reported in few papers. This deleted region has been also described in spindle cell lipoma and (extra) mammary myofibroblastoma, so a morphological and genetic similarity between these three tumor types can be supposed. Moreover, there is the possibility of including cellular angiofibroma, spindle cell lipoma, and (extra) mammary myofibroblastoma in a group of entities that arise from a common stromal precursor cell, which undergoes (myo) fibroblastic or adipocytic differentiation.

Differential Diagnosis

Cellular angiofibroma must be distinguished mainly from angiomyofibroblastoma, spindle cell lipoma, solitary fibrous tumor, and aggressive angiomyxoma.



Spermatic Cord Cellular Angiofibroma, Fig. 1 Cellular angiofibroma presents with a proliferation of bland spindle-shaped cells and numerous irregular, thick-walled blood vessels

Microscopically, angiomyofibroblastoma is very similar to cellular angiofibroma, but it typically shows perivascular accentuation of tumor cells.

Spindle cell lipomas are more cellular than angiofibroma. The stromal collagen of spindle cell lipoma is more brightly eosinophilic and ropey collagen is a characteristic observation in spindle cell lipoma. Moreover, the blood vessels in spindle cell lipoma are usually capillary sized and thin walled.

Distinction should be also made with solitary fibrous tumor that, however, typically shows keloid-type collagen and hemangiopericytoma-like areas. In addition, the rich vascularity with fibrinoid and hyalinized vessels typical of cellular angiofibroma is absent in SFT.

Aggressive angiomixoma occurs most often in the pelvic soft tissues and perineum in young females and it is only exceptionally seen in men. It usually shows an infiltrative growth pattern and invasive borders in contrast to the well-circumscribed lesions of cellular angiofibroma. Furthermore, aggressive angiomyxoma exhibits more numerous blood vessels with large and thick walls and it is positive stained for actin and desmin (usually negative in angiofibroma).

References and Further Reading

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