Case Report

A case of inguinal cellular angiofibroma

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Abbreviations & Acronyms
CAF = cellular angiofibroma
CT = computed tomography
MRI = magnetic resonance imaging
DWI = diffusion weighted imaging

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Introduction: Cellular angiofibroma is a benign mesenchymal tumor that is rare and has a good prognosis. However, preoperative distinction of cellular angiofibroma from malignant tumors is difficult.

Case presentation: A 77-year-old man complained of a left inguinal mass, which was a solid, painless, mobile tumor measuring approximately 40 mm and contacted with the left spermatic cord. Based on his age, the location and imaging findings, a preoperative diagnosis of myxoid liposarcoma was made. The patient underwent left high inguinal orchiectomy with complete resection of the tumor. Histologically and immunohistochemically, the tumor had no feature of malignancy. A postoperative diagnosis of cellular angiofibroma was made. The patient remains free of disease recurrence 12 months after surgery.

Conclusion: Cellular angiofibroma is a benign but rare tumor, which is sometimes difficult to distinguish from malignant neoplasms. Further studies are needed to accurately preoperatively diagnose this tumor.

Key words: cellular angiofibroma, inguinal tumor, myxoid liposarcoma, orchiectomy.

Keynote message

CAF is a benign mesenchymal tumor that is rare and has a good prognosis. A complete excision with negative surgical margins is the recommended strategy. Preoperative distinction of CAF from malignant tumors is difficult.

Introduction

CAF is a benign mesenchymal tumor that is rare. This neoplasm typically develops in subcutaneous tissue of inguinal region in male and the vulvo-vaginal region in female.[1–3] Nucci *et al*. firstly reported CAF of the vulva in female in 1997, and next, Laskin *et al*. reported angiomyofibroblastoma-like tumor of the inguinoscrotal area in male in 1998.[1,2] In accordance with World Health Organization, the terminology "CAF" was applied to tumors of both regions.[4] Patients with CAF present with a gradually enlarging, solid, painless mass.[3] It can easily be misdiagnosed as inguinal hernia in clinical practice. A complete excision with negative surgical margins is the recommended strategy, and long-term follow-up is required.[3] Patients with this neoplasm have a good prognosis. Although a few cases of local recurrence have been reported, there has been no case of metastasis.

We present a 77-year-old male with CAF contacting with the spermatic cord who underwent high inguinal orchiectomy because of the difficulty in preoperatively distinguishing the tumor from a malignant neoplasm, in addition to literature reviews.

Case presentation

A 77-year-old man visited our outpatient department for further examination and treatment of a left inguinal mass discovered incidentally on contrast-enhanced CT. His physical examination revealed a solid, painless, mobile tumor measuring approximately 40 mm in the largest dimension. On CT, the mass contacted with the left spermatic cord and exhibited high

enhancement (Fig. 1). T2-weighted images of MRI demonstrated the tumor had high signal intensity (Fig. 2a), and fat-suppressed T2-weighted images indicated there was no fat tissue in the tumor (Fig. 2b). On DWI, the tumor exhibited diffusion restriction (Fig. 2c). Blood and urine examinations revealed no abnormal findings.

Based on his age, the location and imaging findings, myxoid liposarcoma was suspected. The patient underwent left high inguinal orchiectomy with complete resection of the tumor. Macroscopically, the mass measuring 30 × 25 mm had not infiltrated the testis or the spermatic cord (Fig. 3). Microscopically, the specimen was well circumscribed and exhibited proliferation of spindle-shaped cells and vessels and a mixture of collagen fibers. The cells had no atypism or abnormal mitosis (Fig. 4a). Immunohistochemical examination showed the cells were slightly positive for CD34 (Fig. 4b) and negative for αSMA, desmin, AE1/AE3, S100 protein, STAT6, CDK4, and MDM2. The preoperative diagnosis of myxoid liposarcoma was excluded, and the postoperative diagnosis of CAF was made. The patient remains free of disease recurrence 12 months after surgery.

Discussion

CAF is a rare benign mesenchymal tumor. Its origin remains unknown. Iwasa *et al*. reported that the mean of patient age was 53.5 years (range 22 to 78), and the mass sizes ranged from 0.6 to 25 cm.[3] The tumors often develop in the subcutaneous tissue such as the inguinoscrotal area in male and the vulva in female.[3] However, no sex difference was found in its incidence.[4] Men with CAF are often referred with the chief complaint of a slowly growing, painless inguinal mass. Benign tumors (including fibrous pseudotumor, adenomatoid tumor, and lipoma) and malignant tumors (including liposarcoma, rhabdomyosarcoma, leiomyosarcoma, and paratesticular metastasis) are considered as differential diagnoses in patients who have paratesticular tumors.[5] In particular,

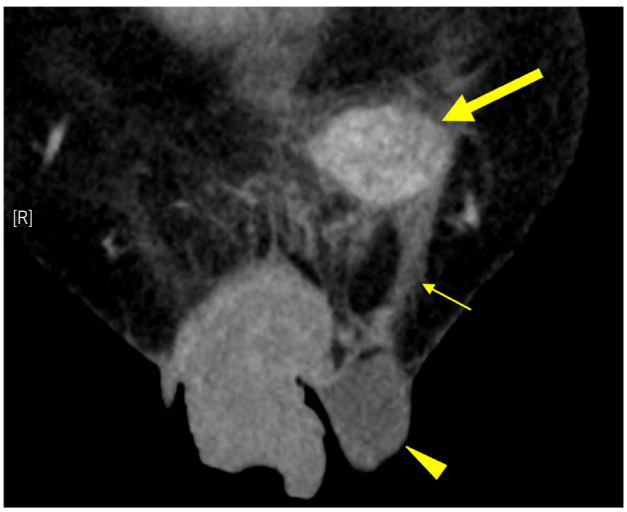

Fig. 1 On contrast-enhanced CT, the arrow indicated a mass that attached to the left spermatic cord (narrow arrow) and exhibited high enhancement. The arrowhead indicated the left testis.

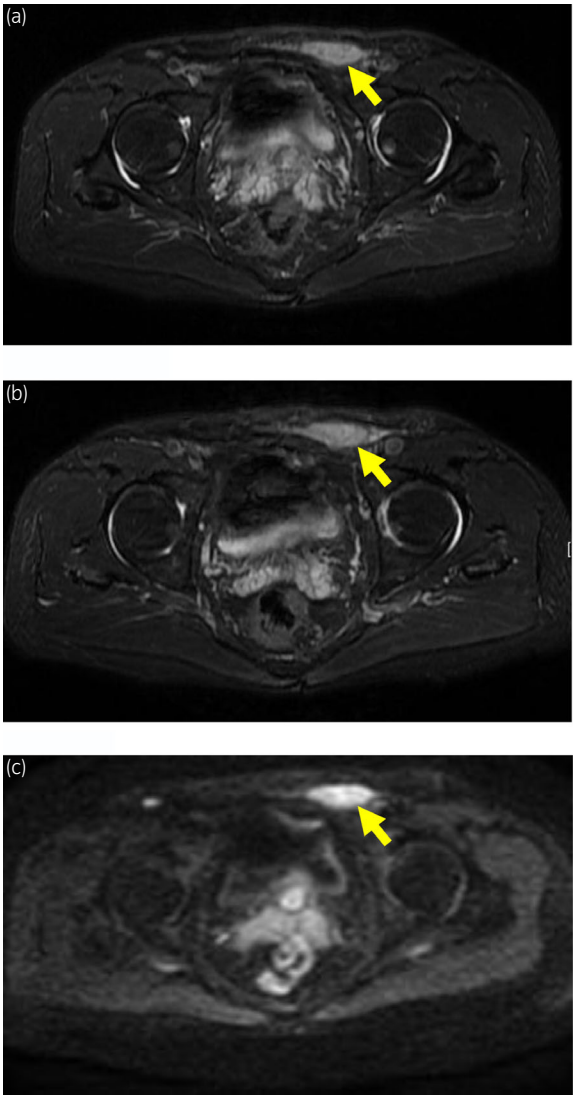

Fig. 2 The arrows indicated the mass attached to the left spermatic cord. (a) On T2-weighted MRI, the tumor had high signal intensity. (b) Fat-suppressed T2-weighted images revealed no fat tissue in the tumor. (c) On DWI, the tumor exhibited diffusion restriction.

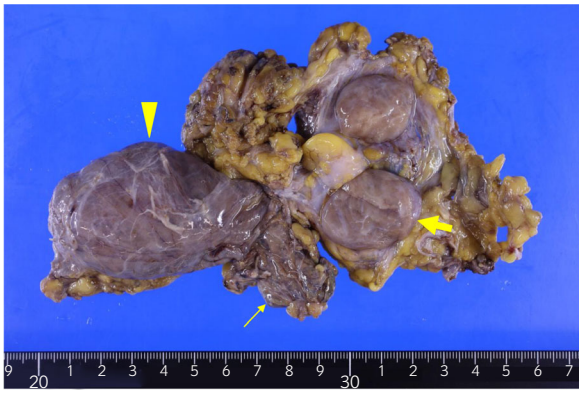

Fig. 3 Macroscopically, the mass (the arrow) measuring 30 × 25 mm had not infiltrated the testis (the arrowhead) or the spermatic cord (the narrow arrow).

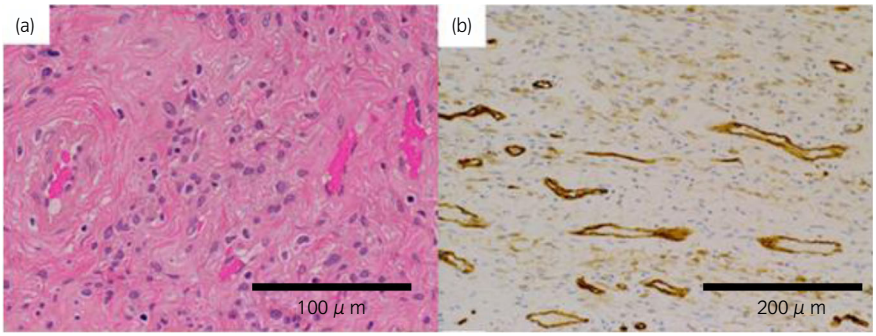

Fig. 4 (a) Histopathological evaluation showed the proliferation of spindle-shaped cells and vessels and a mixture of collagen fibers. The cells did not have atypism or abnormal mitosis (Hematoxylin and eosin). (b) Immunohistochemically (CD34), the cells were slightly positive for CD34.

sarcoma is the most important differential diagnosis in clinical practice because wide resection is recommended for these malignant tumors.[6]

Histologically, CAF is generally a well-marginated tumor, including short bundles of collagen, spindle-shaped cells, and many small- to middle-sized round vessels.[1–3] Generally, significant pleomorphism and abnormal mitoses are not observed. Immunohistochemically, Iwasa *et al*. demonstrated that 60% of the tumors were positive for CD34, suggesting vascular origin, and a minority of tumors were positive for SMA and desmin. No tumors were positive for S100 protein. In female cases, the tumors were often positive for estrogen receptor and progesterone receptor.[3] In our patient, the histological and immunohistochemical characteristics corresponded with those previously reported.

On imaging, some findings depending on the histological appearance have been noted.[5] On MRI, CAF demonstrates heterogeneous increased signal intensity on T2-weighted imaging, and heterogeneous contrast-enhanced pattern due to hypervascularity. Although gadolinium was not administered, signal intensity of the tumor on T2-weighted imaging was observed in our patient. Intratumoral fat was reported to be presented in 24–56% of cases.[1,3,6] Miyajima *et al*. reported that the presence of a well-marginated hypervascular mass containing fat in a male inguinal region suggested CAF.[6] Moreover, CAF was revealed to have no area of diffusion restriction on DWI, which is mainly noted in malignant tumors.[7,8] These characteristics are different from those of our patient. Therefore, MRI findings are non-specific, and preoperative distinction of CAF from malignant tumors is difficult.

Myxoid liposarcoma, one subtype of liposarcoma, considered as a preoperative diagnosis in our case, exhibits similar findings to CAF on MRI. The tumor often has only minimal fat content and may not exhibit the signal intensity of fatty tumors.[9] Furthermore, due to the abundance of water, the tumor exhibits high intensity on T2-weighted images with fat suppression.[9] In our patient, the tumor had similar characteristics and was misdiagnosed as a malignant neoplasm. In contrast, several reports demonstrated that on dynamic contrast-enhanced MRI imaging, benign tumors have an initial upstroke of signal intensity, followed by either plateaus or gradual increases,[10] whereas malignant tumors have an initial upstroke, and subsequent gradual washout.[10] Thus, dynamic contrast enhancement may be useful to distinguish CAF from malignant tumors.

The recommended strategy is complete excision with negative surgical margins. Iwasa *et al*. reported that patients who underwent complete tumor excision had a good prognosis and no recurrence.[3] In contrast, a few cases who experienced local recurrence have been reported.[2,11] Thus, long-term follow-up may be required.

Conflict of interest

The authors declare no conflict of interest.

References

1 Nucci MR, Granter SR, Fletcher CD *et al*. Cellular angiofibroma: a benign neoplasm distinct from angiomyofibroblastoma and spindle cell lipoma. *Am. J. Surg. Pathol.* 1997; **21**: 636–44.
2 Laskin WB, Fetsch JF, Mostofi FK *et al*. Angiomyofibroblastoma-like tumor of the male genital tract: analysis of 11 cases with comparison to female angiomyofibroblastoma and spindle cell lipoma. *Am. J. Surg. Pathol.* 1998; **22**: 6–16.
3 Iwasa Y, Fletcher CD. Cellular angiofibroma: clinicopathologic and immunohistochemical analysis of 51 cases. *Am. J. .Surg. Pathol.* 2004; **28**: 1426–35.
4 Fletcher CDM, Unni KK, Mertens F (eds). *World Health Organization Classification of Tumours: Pathology and Genetics of Tumours of Soft Tissue and Bone*. IARC Press, Lyon, France, 2002.
5 Mathur M, Spektor M. MR imaging of the testicular and extratesticular tumors: when do we need? *Magn. Reson. Imaging Clin. N. Am.* 2019; **27**: 151–71.
6 Miyajima K, Hasegawa S, Oda Y *et al*. Angiomyofibroblastoma-like tumor (cellular angiofibroma) in the male inguinal region. *Radiat. Med.* 2007; **25**: 173–7.
7 Ntorkou AA, Tsili AC, Giannakis D *et al*. Magnetic resonance imaging findings of cellular angiofibroma of the tunica vaginalis of the testis: a case report. *J. Med. Case Rep.* 2016; **10**: 71.
8 Maruyama M, Yoshizako T, Kitagaki H *et al*. Magnetic resonance imaging features of angiomyofibroblastoma-like tumor of the scrotum with pathologic correlates. *Clin. Imaging* 2012; **36**: 632–5.
9 Abete L, Simonato A, Toncini C *et al*. Myxoid liposarcoma of the spermatic cord: US and MR imaging findings. *J. Clin. Ultrasound* 2014; **42**: 96–9.
10 Tsili AC, Argyropoulou MI, Astrakas LG *et al*. Dynamic contrast-enhanced subtraction MRI for characterizing intratesticular mass lesions. *AJR Am. J. Roentgenol.* 2013; **200**: 578–85.
11 McCluggage WG, Perenyei M, Irwin ST *et al*. Recurrent cellular angiofibroma of the vulva. *J. Clin. Pathol.* 2002; **55**: 477–9.