

Case Report

A rare mesenchymal neoplasm at unusual location: Solitary fibrous tumor of vulva



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Introduction

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm and accounts for less than 2% of all soft tissue tumors. Moreover, the genital tract location of this tumor is even rarer, with just 5 cases of vulval SFT reported in literature (Taki et al., 2012). We recently managed a case of SFT of vulva which was diagnosed initially as dermatofibrosarcoma protuberans. In this article we intend to discuss this case and review the limited available literature for various presentations, diagnosis and treatment outcomes of vulval SFT.

Presentation of case

A 57 year old female presented with complaints of painless swelling over the vulva which has been slowly increasing in size over the past 10 years. There was no history of fever, weight loss, loss of appetite, discharge, or pigmentation. A biopsy performed elsewhere previously, had diagnosed the current lesion as 'dermatofibrosarcoma protuberans' (DFS). Past history was significant for similar lesion at the same location which was excised about twenty years ago. There was no past history of tuberculosis including her family.

The patient was moderately built and nourished (BMI: 25 kg/m²). Local examination of vulva revealed a large (8 by 10 cm) irregular lesion, present over the left labia majora and mons pubis (Fig. 1). The lesion had appearance of multiple confluent nodular lesions each measuring about 0.5 to 2 cm. On palpation the lesion was firm to hard, indurated and non-tender with clear margins. The lesion was noticed to cross the midline involving clitoris. There were no palpable inguinal or supraclavicular nodes.

Ultrasonography revealed a lobular solid tumor in the subcutaneous plane. Tumor markers (CA-125, carcinoembryonic antigen, desmin and epithelial membrane antigen) were negative. She underwent wide excision, ensuring clear margins using frozen section procedure. This was followed by skin grafting from the thigh (Fig. 1). Histopathology of the excised lesion was reported as SFT. The tumor was smooth and white on cross-section. The microscopic picture was that of a pattern less fibroblastic spindle cells in a hyalinized collagen matrix (Fig. 2). On immunohistochemical staining, the tumor was positive for CD34 and bcl-2, but negative for S-100, a SMA, EMA and AE1/AE3. No adjuvant therapy was administered and the patient made an un-eventful recovery, with no recurrence so far for 24 months.

Discussion

The case discussed is that of an unusual presentation of a rare neoplasm of soft tissue – the SFT. This tumor was first described arising from the pleura in 1931 (Hasegawa et al., 1999), which is the most common site. Since then, it has been described at locations such as the meninges, peritoneum, liver, upper respiratory tract, orbit, thyroid, and salivary gland (Hasegawa et al., 1999). Gynecological SFT, however, is a rare entity (Taki et al., 2012). Of all the reports on gynecological SFTs, the vulval origin has been described in only five reports (Table 1) (Taki et al., 2012; Fukunaga, 2000; Nielsen et al., 1997; Biedrzycki et al., 2007; He et al., 2010).

The age of our patient at presentation was similar to the age reported in literature for gynecological SFT, which is an average of 50 years, ranging from 14 years to 78 years. The presentation is that of a slowly growing painless lump. The tumor in this case grew over 10 years to a significant size of 8 × 10 cm. This slow growth along with the absence of nuclear atypia and metastasis suggests benign nature.

Histologically, it is characterized by fibroblastic spindle cell proliferation arranged in no particular pattern in a collagenous stroma. The important differentials for this tumor are the spindle cell tumors, tumors of neural origin, smooth muscle tumors, hemangiopericytoma (HPC), DFS,

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Fig. 1. (A) Vulval solitary fibrous tumor 8 × 10 cm involving clitoris, (B) Wide excision and skin grafting.

and spindle cell lipoma (SCL) (Kanagaraja et al., 2013). In the case discussed, the tumor was reported initially to be a DFS; however it was found later to be a SFT. Both SFT and DFS express CD34 reactivity. However, hyalinization was noticed in the present tumor and this is not a feature of DFS; moreover, features of prominent storiform pattern and xanthomatous cells seen in DFS were absent. Immunohistochemical detection of CD34 and bcl-2 was helpful in differentiating SFT from other spindle-cell tumors (Hasegawa et al., 1998). Immunostaining for S-100 can exclude tumors of neural origin. Smooth muscle tumors show lesional reactivity for smooth muscle actin, while SFT only shows reactivity in vessel walls. This reactivity pattern of SFT is not seen in HPC, and hence helps in differentiation. Spindle cell lipoma can be differentiated by the presence of mature fat cells which were absent in this case. In SFT, certain pathological features predict aggressive and malignant behavior and hence are important to look for, as these have prognostic and treatment implications. None of these features were seen in our case or in any of the reported cases of vulval SFT (Taki et al., 2012; Fukunaga, 2000; Nielsen et al., 1997; Biedrzycki et al., 2007; He et al., 2010). These pathological features include – nuclear atypia, hypercellularity, frequent mitoses, necrosis and infiltrative growth. Besides these, immunohistochemical staining positivity with α -SMA and S-100 has been shown to predict malignant behavior (Vallat-Decouvelaere et al., 1998). Adjuvant radiotherapy is often performed following excision in these cases.

The tumor involves the adjacent tissues requiring significant excision, has recurrences and there is at least one case report of metastasis (Vallat-Decouvelaere et al., 1998). Hence, despite benign behavior, early aggressive management with wide excision ensuring clear margins is recommended. Wide excisions do have significant functional impairment and hence a precise pathological studies and imaging are

necessary. Due to its superficial location ultrasound was done to assess the nature and extent of the mass. There are case reports of excessive bleeding associated with resection and hence particularly in deeper SFTs, contrast enhanced imaging to assess the vascularity has been recommended. Due to recurrent nature, a long term follow-up must be advised. Unfortunately, for vulval SFT, there is no data on long term follow-up. Our case report has longest reported follow-up period for vulval SFT. Similar to our case, there has been no reported recurrence of vulval SFT prospectively, so far. Recurrence, however, is a known phenomenon for gynecological SFTs in other locations (Vallat-Decouvelaere et al., 1998). Our case had a history of similar presentation; hence the current lesion may be a recurrence. However, in the absence of histopathological evidence, this is difficult to substantiate.

Conclusion

In conclusion, the clinicians and pathologists should be aware of this rare differential of vulval mass. Although usually benign, the tumor requires thorough pathological and radiological workup to rule out malignant features and other close differentials. Early aggressive resection is recommended due to destructive nature of this tumor on adjacent tissues. The tumor has a tendency for recurrence and hence the resected margins should be clear and long term follow-up should be arranged. Although the experience with follow-up of this tumor is scarce, the prognosis depends on complete surgical resection and lack of pathological features of malignancy.

Conflict of interest

The authors have no conflict of Interest to report.
Informed consent was obtained for the purpose of this case report.

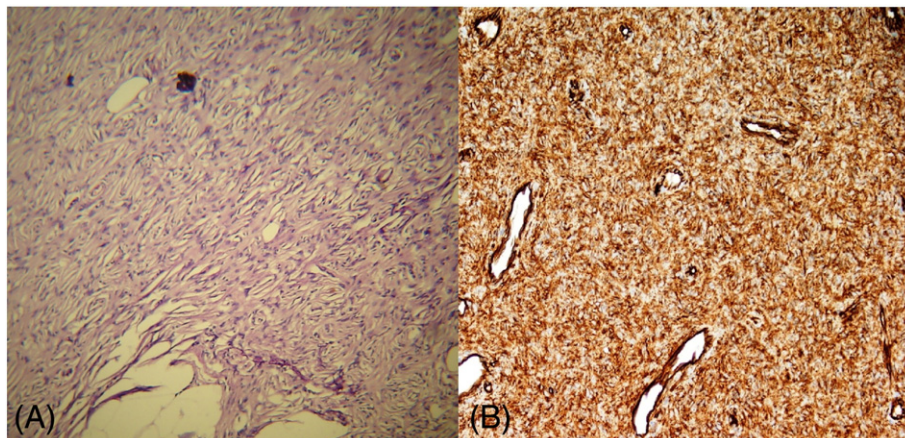


Fig. 2. (A) Hematoxylin and eosin stained section ($\times 100$) showing patternless arrangement of spindle shaped cells with partially hyalinized matrix. (B) Intense CD34 staining.

Table 1
Case reports of vulval solitary fibrous tumor.

Author	Age (Years)	Size and presentation	Immuno-histochemical staining	Outcome ^a
Taki et al. (2012)	56	5 cm; no symptoms	CD34 ⁺ ve, and AE1/AE3, EMA, S-100, a-SMA ⁻ ve.	No recurrence for at least 18 months
Fukunaga (2000)	70	15 cm; 15 years of slow growing mass	Vimentin, CD34, progesterone receptors, and bcl-2 ⁺ ve.	No recurrence for at least 9 months
Nielsen et al. (1997)	51	5 cm; painless slowly growing lump	Vimentin, CD34 ⁺ ve, and cytokeratin, desmin, S-100 protein, EMA, SMA ⁻ ve.	No recurrence for at least 12 months
Biedrzycki et al. (2007)	45	6 cm; painless slowly growing lump	CD34, Bcl 2, and vimentin, CD99 ⁺ ve, MNF116, desmin, SMA, ER, PR, and S100 ⁻ ve.	No recurrence for at least 6 months
He et al. (2010)	39	10 cm; slowly growing painless lump	CD99, vimentin, CD34 ⁺ ve, S100, SMA, desmin, ER, PR ⁻ ve.	No recurrence for at least 10 months

^a All cases involved wide excision.

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