Case Report

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Pathology Section

A Rare Mesenchymal Neoplasm at an Uncommon Anatomical Site-Solitary Fibrous Tumour of Vulva

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ABSTRACT

Previously, Solitary Fibrous Tumour (SFT) was thought to originate from the pleura, a relatively rare mesenchymal neoplasm with an indolent course. Vulva is a very uncommon site of extrathoracic SFT. This is a report of a very uncommon case of vulval SFT in a 30-year-old female with complaint of a swelling over vulval region. Fine Needle Aspiration Cytology (FNAC) of the growth was attempted twice and only blood was aspirated. The swelling was excised completely. On histological examination, the sections revealed a cellular tumour of ovoid-to-fusiform spindle cells with indistinct cell borders, with haphazard arrangement or patternless pattern along with proliferation of variable sized blood vessels. The diagnosis was finalised to be a benign stromal tumour (most probably SFT), which was confirmed on immunohistochemical examination using Cluster of Differentiation 34 (CD34), CD99 and Signal Transducer and Activator of Transcription 6 (STAT6) markers.

Keywords: Extrathoracic, Haemangiopericytoma, Signal transducer and activator of transcription 6

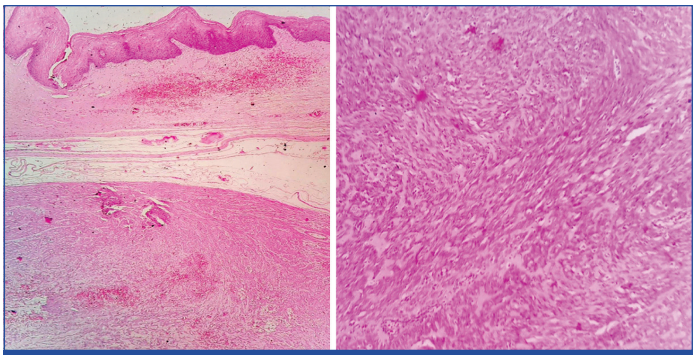
CASE REPORT

A 30-year-old married female presented to the Obstetrics and Gynaecology outpatient department, with the history of swelling over vulval region since 14 months. The swelling was gradual in onset and progressively increasing in size. There was no significant family history, no history of intake of oral contraceptive pills or any medication with normal menstrual cycle.

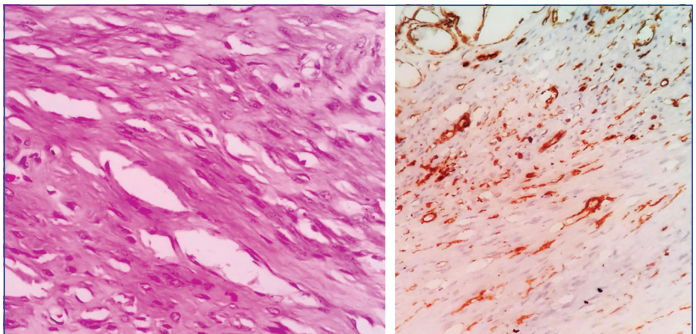
On local physical examination, the swelling was skin-fixed, non tender, mobile and soft to firm in consistency with cauliflower like growth on surface. The FNAC of the swelling, along with pap smear was advised to the patient. The FNAC was attempted twice and only blood was aspirated. Pap smear revealed superficial and intermediate squamous cells in sheets with no parabasal and basal cells. Few intermediate cells showed koilocytic changes in an acute inflammatory background. No endocervical cells were seen. No epithelial cells abnormality was seen. As per the Bethesda system of reporting of cervical cytology, pap smear was negative for intraepithelial lesion and malignancy, with presence of acute inflammation [Table/Fig-1] [1]. Ultrasonography (USG) of lower abdomen revealed uterus and bilateral tube and ovary to be within normal limit. The cervix appeared bulky measuring 3 to 7 cm in size with minimal free fluid in cul-de-sac. The USG findings were suggestive of cervicitis with minimal free fluid in cul-de-sac.

The swelling was excised and sent for histopathological examination. On gross examination, the specimen was a greyish-white skin covered irregular piece of tissue with multiple finger like projection measuring 3.0×3.0×1.5 cm. The cut surface showed a well circumscribed greyish white nodule beneath the skin [Table/Fig-2]. Multiple tissue sections were processed to obtain 3 to 4 µm thick section, and stained with Hematoxylin and Eosin (H&E). On histopathological examination, the sections revealed papilliferous hyperplastic squamous epithelium; tumour nodule lying in subepithelium showing proliferation of fibroblastic cells; ovoid to fusiform spindle cells with indistinct cell borders, with haphazard arrangement or patternless pattern along with proliferation of variable sized blood vessels [Table/Fig-3-5]. Overall, histological

[Table/Fig-1]: Showing superficial and intermediate squamous cells in sheets with no parabasal and basal cells seen in an acute inflammatory background (Pap smear, 40X); **[Table/Fig-2]:** Cut surface showing well circumscribed nodule with grey-white surface and overlying skin surface yellowish white papillary projection. (Images from left to right)

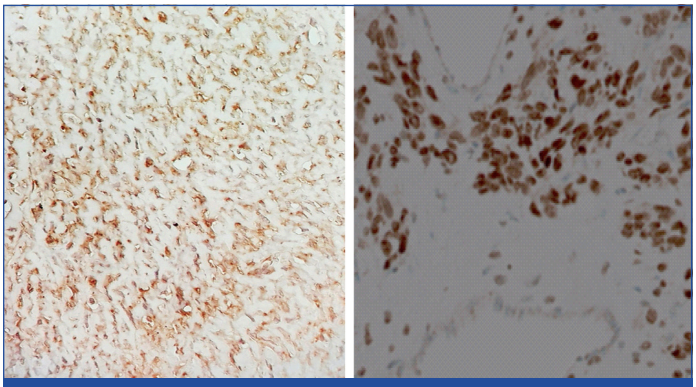


[Table/Fig-3]: Showing epidermis, haemorrhagic area and well-circumscribed tumour present in sub epidermal region showing proliferation of spindle cells in haphazard pattern with variable sized slit like spaces (H&E, 4X). **[Table/Fig-4]:** Showing proliferation of fibroblastic cells; ovoid to fusiform spindle cells with indistinct cell borders, with haphazard arrangement or patternless pattern along with proliferation of variable sized blood vessels (H&E, 10X). (Images from left to right)



[Table/Fig-5]: Showing ovoid to fusiform spindle cells with indistinct cell borders along with proliferation of variable sized blood vessels (H&E, 40X).
[Table/Fig-6]: Showing CD34 positivity of blood vessels and tumour cells (IHC, 40X). (Images from left to right)

features were suggestive of benign stromal tumour favouring SFT. On immunohistochemistry (IHC), the tumour was found to be positive for CD34, CD99 and STAT6 markers [Table/Fig-6-8]. On the basis immunohistochemistry the diagnosis of SFT was confirmed. The patient made an uneventful recovery, and was on regular follow-up. There has been no recurrence so far for nine months. No adjuvant therapy was given.



[Table/Fig-7]: Showing CD99 positivity in tumour cells (IHC, 40X).
[Table/Fig-8]: Showing nuclear positivity with STAT6, (IHC, 40X). (Images from left to right)

DISCUSSION

The SFT is a relatively rare neoplasm of mesenchymal origin, previously described as originating from the pleura as mesothelial tumour. But presently it is considered to originate from fibroblasts in any part of the body and is known to have an indolent course [2]. However, there are reports of extrathoracic SFTs occurring in soft tissues like extremities and genital tract. This tumour was first described in 1931 arising from the pleura, which is the most common site. Since then, there were reports of its occurrences in meninges, peritoneum, liver, upper respiratory tract, orbit, thyroid and salivary gland [3-6]. Gynaecological SFT is a rare entity and the vulva has been the very uncommon site. Another subset of SFT, hemangiopericytoma is distinguished from the former by its branching vascular pattern. There is no gender predominance for SFT [4]. The SFT is a fibroblastic tumour that shows positivity for CD34, CD99 and STAT6. Several translocations have been found in SFT including t (12:19), and t (3:12) chromosome sites [5]. The fusion gene arises from recurrent intrachromosomal rearrangement on 12q13 chromosome [5].

Morphologically, SFT is characterised by proliferation of spindle cells in blends of hyper and hypocellular area. Haemangiopericytoma, a histological type of SFT shows thin walled branching vascular pattern termed as "staghorn" appearance with a prominent pericystic pattern [7]. Other histological variant includes lipomatous SFT; a rare variant containing variable amount of mature fat admixed in tumour part and SFT with giant cells showing classical feature of SFT along with pseudovascular space lined by multinucleated stromal giant cells [5]. Among all cases of gynaecological SFT reported, the vulval origin has been described in nine case reports, so far [Table/Fig-9] [2,4,6,8-13]

The important differential diagnosis of SFT includes other spindle cell tumours like Fibrous Histiocytoma (FH), Synovial Sarcoma (SS) and mesenchymal chondrosarcoma, tumour of neuronal origin, smooth muscle tumour and spindle cell lipoma [5,8]. The FH shows more prominent and uniform spindle cell pattern with storiform arrangement which was not seen in the present case, hence differentiating it from SFT. About 10 to 20% cases of SS shows focal haemangiopericytoma like pattern but almost always associated with distinct spindle cells, hyalinised-calcified areas, glands and expression of cytokeratin, while CD34 expression is not seen [5]. However, hyalinised-calcified area was absent and CD34 expression was seen in the present case. Immunohistochemical positivity of CD34 and B-cell Lymphoma 2 (bcl-2) differentiates it from other spindle cell tumours [14]. Smooth muscle tumours show smooth muscle actin positivity, while only vessel walls positivity in SFT. This reactivity pattern is not seen in hemangiopericytoma, hence differentiating it from SFT. Immunostaining with S100 excludes tumours of neural origin. Spindle cell lipoma can be differentiated by the presence of mature fat cells from SFT, while lipomatous SFT is difficult to differentiate from spindle cell lipoma which can be distinguished based on Immunohistochemistry (IHC) [8]. Aggressive angiomyxoma is another differential diagnosis, which is relatively hypocellular tumour, consisting stellate fibroblasts in a prominent myxoid stroma with interspersed blood vessels [6].

Author, year and place of the study	Age (years)	Size and presentation	Outcome
Nielsen GP et al., 1997, Boston [11]	51	5 cm; painless slowly growing mass	No recurrence for until 12 months
Fukunaga M, 2000, Tokyo, Japan [12]	70	15 cm; 15 years of slow growing mass	No recurrence for until 9 months
Biedrzycki OJ et al., 2007, London, UK [13]	45	6 cm; painless slowly growing mass	No recurrence for until 6 months
He Y et al., 2010, Chengdu, China [9]	39	10 cm; painless slowly growing mass	No recurrence for until 10 months
Taki M et al., 2012, Kyoto, Japan [10]	56	5 cm; no symptoms	No recurrence for until 18 months
Burnett LA et al., 2012, Pittsburgh, USA [4]	60	5 cm; 4 months of mild pain with slowly growing mass	No recurrence for until 30 months
Nag G and Rao SR, 2015, Karnataka, India [8]	57	8 cm; 10 years of painless low growing mass	No recurrence for until 24 months
Pearee DC et al., 2017, Baltimore, USA [2]	64	9.8 cm; 15 years of painless slow growing mass	Patient died, 15 months from the original diagnosis of SFT and approximately 10 months after spinal metastasis
Rekhi B et al., 2021, Mumbai, India [6]	24	10 cm; 3 year of slow growing mass	-
Present case report, Uttar Pradesh, India	30	3 cm; 14 months of painless slow growing mass	No recurrence for until 9 months

[Table/Fig-9]: Compilation of published case reports on vulval solitary fibrous tumour. [2,4,6,8-13].

Accurate characterisation and identification of the lesion need to further stratify a patient's prognosis on the basis of variations in morphology and immunohistochemical profile of an excised tumour so that treatment strategy can be changed accordingly. The literature has demonstrated both malignant and benign case studies of these tumours and has attempted to identify the specific characteristics for malignant and life threatening tumours [4].

The subsequent pathological characteristics need to be included for identification of malignant behaviour of lesion such as: nuclear atypia, hypercellularity, frequent mitoses greater than 4/10 High-power Fields (HPF), necrosis and infiltrative growth. Besides these, Alpha-smooth Muscle Actin (α-SMA) and S100 positivity has been shown to predict malignant behaviour [15]. The tumours involving the adjacent tissues require significant excision at least margins should be free of tumour due to risk of recurrence and metastasis. The tumour can recurred locally and metastasise to genitourinary tract and spine. Adjuvant chemotherapy should be performed after excision if high risk findings are demonstrated in patient tumour particularly high level of mitotic figure and should not be given to index patient because of rarity of malignant transformation [8]. In evaluation of adjuvant chemotherapy, the data has been contradictory for temozolomide with bevacizumab showing significant antitumour activity by some studies while others demonstrating no benefit compared to single agent temozolomide or dacarbazine.

CONCLUSION(S)

Vulva is a very uncommon site of gynaecological SFT. The present case is a 10th rare case report of few vulvar SFTs. The SFT was diagnosed on histological examination; however, diagnosis should be confirmed by immunostaining using CD34, CD99 and STAT6 markers. The NGFI-A Binding protein 2 and Signal Transducer and Activator of Transcription 6 (NAB2-STAT6) fusion gene was used to achieve exact diagnosis for rare site of SFT.

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