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MYXOID DERMATOFIBROSARCOMA PROTUBERANS OF THE VULVA WITH MYOID NODULES: CLINICOPATHOLOGIC AND IMMUNOHISTOCHEMICAL STUDY OF A CASE

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Abstract
Dermatofibrosarcoma protuberans is a slow growing dermal spindle cell tumor seldom seen in the vulva and its myxoid variant, a rare type of dermatofibrosarcoma protuberans is characterised by extensive myxoid degeneration. We present the case of a 62 year old woman with an enlarging vulval swelling. Mass was excised surgically. Histopathologically the tumor consisted of uniform spindle-shaped cells showing strong positivity with CD34. In addition to the typical storiform pattern and lace like infiltration, prominent myxoid stromal changes were seen. Herein we report an interesting case of myxoid dermatofibrosarcoma protuberans, uncommonly reported in the dermatopathology literature.

Key words: dermatofibrosarcoma protuberans; myoid nodules; myxoid variant; vulva

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Introduction

Dermatofibrosarcoma protuberans (DFSP) is a rare, locally aggressive dermal mesenchymal neoplasm that usually occurs on the trunk and extremities [1]. The characteristic histologic feature of DFSP is the proliferation of densely packed monomorphous spindle cells, arranged in a storiform pattern. It is uncommon in vulva and usually unsuspected clinically. To our knowledge, only 29 cases of DFSP of vulva have been reported thus far [2]. Herein we present an additional case of this tumor with myxoid areas and discuss its clinicopathological and immunohistochemical features.

Case Report

A 62 year old woman presented with an enlarging vulval swelling. The lesion had been growing over a period of 6 months. There was no history of weight loss, loss of appetite, vaginal or rectal bleeding, pain, fever, itching, trauma or surgery in the affected area. Five years earlier, the patient had undergone hysterectomy for uterine leiomyoma.

Clinical examination revealed a hard, bosselated, mobile mass measuring 6 x 8 cm, extending medially to labia minora. Mass was excised surgically and subjected to histopathological examination.

The gross specimen consisted of skin covered swelling measuring 10 x 7 x 5 cm. On sectioning the surface showed homogeneous glistening white, vaguely lobulated areas with myxoid areas (Fig. 1).

Microscopy showed hyperkeratotic epidermis,a subepidermal grenz zone and a partially circumscribed cellular spindle cell tumor composed of spindle shaped fibroblast like cells arranged in repetitive short intersecting fascicles (Fig. 2), imparting a storiform configuration with a very occasional mitosis (1/10 hpf) along with extensive foci of myxoid change containing prominent thin walled vasculature focally showing myoid nodules (Fig. 3). Tumor was seen infiltrating the adjoining adipose tissue in a characteristic lace like pattern with involvement of all margins and base.

Immunohistochemically, the tumor cells showed strong positivity for CD34. No staining of tumor cells with S-100 and Smooth muscle actin was seen leading to a diagnosis of myxoid dermatofibrosarcoma protuberans with myoid nodules.

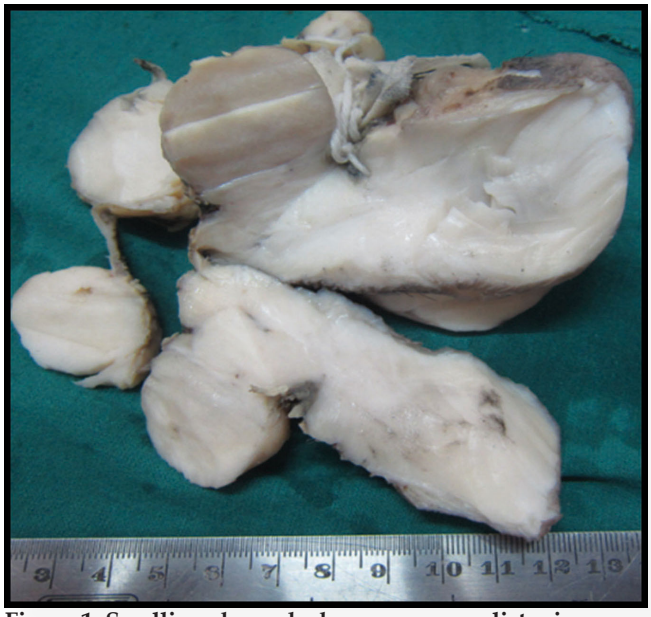

Figure 1. Swelling showed a homogeneous glistening white,vaguely lobulated cut section

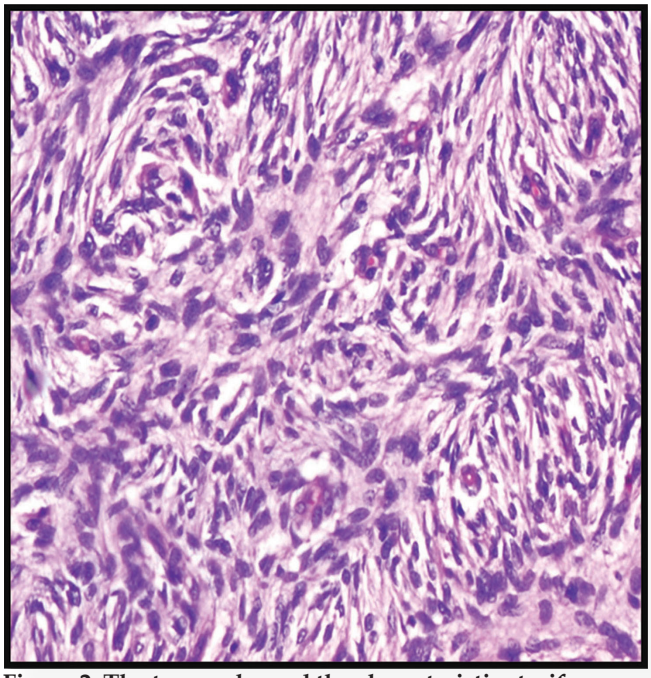

Figure 2. The tumor showed the characteristic storiform pattern (H&E, x 40)

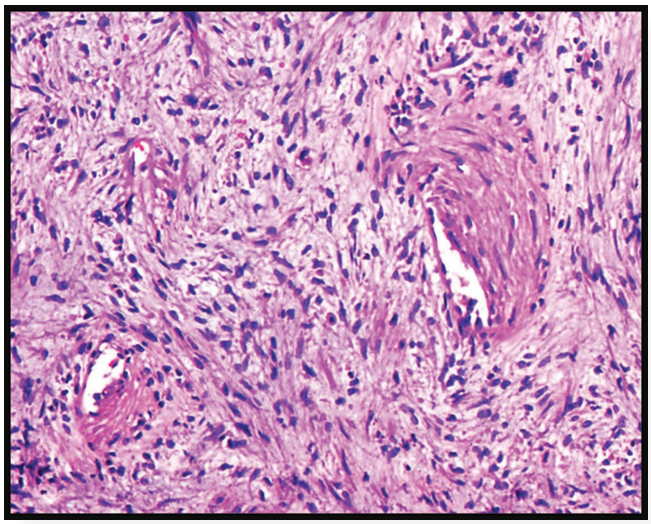

Figure 3. Myoid nodule centred along a blood vessel (H&E, x 40)

Discission

DFSP is a rare, slow growing, but locally aggressive tumor. DFSP of the vulva typically occurs as a firm, well circumscribed nodular mass attached to the overlying skin, but movable over the deeper tissues. Occurrence at sites of previous trauma has been reported, and many patients have a previous, long preoperative history [3].

Our patient neither had previous trauma nor operation history, except hysterectomy for uterine leiomyoma, 5 years ago. Histologically, DFSP consists of relatively uniform spindle cells containing elongated nuclei, without significant cytologic atypia or pleomorphism, and arranged in a predominantly typical storiform pattern. Although the tumor is usually located in the dermis, it invariably shows the infiltrative growth pattern, with trapping of the subcutaneous fat tissue in the characteristic honeycomb appearance.

Myxoid DFSP is a rare variant of this neoplasm, characterized by prominent myxoid stromal changes. The first such case was cited in 1983 by Frierson and Cooper [4]. Since then only a few cases have been reported. The pathogenesis of myxoid change remains uncertain, with majority of the cases presenting as a slowly growing, firm subcutaneous mass. The commonly involved sites were the extremities, followed by the head and neck [5]. In addition to the typical histological features of DFSP, the tumor cells in the myxoid type of DFSP are embedded in an abundant, palely eosinophilic myxoid stroma with prominent thin-walled vessels, the latter are frequently present throughout the tumor [5]. An unusual feature of DFSP is the myoid nodule. Originally construed as evidence of myofibroblastic differentiation [6], these structures seem to be centred in some cases around blood vessels [7,8], as seen in our case and likely represent an unusual non neoplastic vascular response to the tumor. Immunohistochemical findings are consistent with the typical DFSP, with the positive staining for CD34 ranging from 84% to 100%, and negative for other markers, such as S-100, desmin and actin [9].

Prominent myxoid changes can often obscure the typical storiform pattern, causing considerable diagnostic confusion [10]. The differential diagnoses of DFSP are diverse and the entities that may be considered are shown below in a tabular form Table I.

In summary, we present an interesting case of Myxoid DFSP, uncommonly reported in the dermatopathology literature. Possibility of this variant of DFSP should also be considered while evaluating myxoid soft tissue neoplasms.

Entity	Distinguishing features
Benign fibrous histiocytoma	- Short fascicles, haphazard growth pattern. - Presence of secondary element (giant cells, inflammatory cells). - **CD34**: Focal staining in occasional cases.
Myxoid neurofibroma	- Presence of tactoid structures, wavy buckled nuclei. - Lack of highly cellular areas with mitotic figures. - **S-100**: Protein positivity.
Myxoid liposarcoma	- Presence of lipoblasts and atypical, undifferentiated mesenchymal tumor cells. - **CD34:** Usually negative.
Myxofibrosarcoma	- Well-circumscribed tumor, with overall increased cellularity. - **EMA:** Positive staining in many low grade myxofibrosarcoma.
Superficial angiomyxoma	- Lesions tend to be displayed in a lobular growth pattern. - Scattered neutrophils surrounding the vessels.

Table I. The differential diagnoses of Dermatofibrosarcoma protuberans

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