Recurrent Dermatofibrosarcoma Protuberans with Pigmentation and Myoid Differentiation

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Case Report

A 44-year-old woman presented to the Lok Nayak Jai Prakash Hospital, New Delhi, India, in 2017 with a recurrent mass on her right upper arm. The tumour had regrown since being surgically removed three years prior, at which time it had been histopathologically diagnosed as a DFSP. At presentation, the lesion was attached to the overlying skin, measuring 8 x 7 x 6 cm. A computed tomography (CT) scan showed a homogeneously-enhancing soft tissue lesion in the subcutaneous plane of the lateral aspect of the middle-third of the right arm, measuring 7 x 6 x 4.5 cm [Figure 1A]. The lesion had well-defined margins except medially, where it was abutting the deltoid muscle. There was no evidence of organomegaly or lymphadenopathy. The patient subsequently underwent wide surgical excision of the lesion.

Upon gross examination, the excised soft tissue mass measured 13 x 11 x 5 cm. The surface of the lesion showed a firm homogenous greyish-white growth of 6 x 6 x 4.5 cm, which extended deep into the underlying dermis [Figure 1B]. There were no areas of haemorrhage or necrosis. Histologically, the overlying skin was unremarkable. The tumour itself was located in the dermis and was composed of monomorphic spindle cells arranged in a storiform and fascicular pattern with a parallel arrangement of cells [Figure 2A]. In certain areas, the tumour was more cellular and composed of elongated cells with moderate cytoplasm and elongated-to-plump nuclei, which were mostly perivascular [Figure 2B]. The eosinophilic cells were

Dermatofibrosarcomas (DFSP) are rare low-grade sarcomas involving the dermis and subcutaneous tissue.1,2 This type of tumour has a low rate of distant metastasis, but a higher rate of local recurrence. Histological subtypes of DFSP include giant-cell, pigmented, sclerosing, atrophic, myoid and fibrosarcomatous types.1,2 Of these, pigmented DFSP are characterised by interspersed melanin-rich dendritic cells and are very rare, accounting for less than 5% of all reported cases of DFSP.1 However, the origin of myoid differentiation in DFSP and its clinical significance is unknown. This case report describes a case of myoid differentiation in recurrent pigmented DFSP presenting as an arm swelling in an adult female patient.

Abstract: Dermatofibrosarcomas (DFSP) are rare low-grade tumours with various subtypes and usually occur among middle-aged adults. However, myoid differentiation is very rare. We report a 44-year-old woman who presented to the Lok Nayak Jai Prakash Hospital, New Delhi, India, in 2017 with a recurrent pigmented DFSP presenting as an arm swelling. Upon histological and immunohistochemical analysis, myoid differentiation was confirmed. A literature review of the clinical and histopathological features of this rare entity is presented.

Keywords: Dermatofibrosarcoma Protuberans; Melanocytes; Pigmentation; Cell Differentiation; Case Report; India.
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Discussion

In general, DFSP occur mostly in middle-aged adults, but have been reported in patients between 8–87 years old. Men are affected more often than women at a ratio of 2.3:1. This type of tumour is usually painless and presents as a long-standing solitary multinodular mass. Commonly affected sites include the trunk, particularly the back, and the limbs, chest and head and neck region. The tumour is locally aggressive and usually infiltrates the deep dermis and subcutaneous fat. A genetic study found that 89% of DFSP cases had collagen type I alpha 1 platelet-derived growth factor beta fusion transcripts. A classical histological feature of DFSP is a storiform growth pattern of benign-looking spindle cells. Histopathologically, CD34 is considered a highly-specific marker of DFSP.

The origin of muscle differentiation in DFSP is debatable. Calonje et al. first described myoid differentiation in five cases of DFSP either with or without fibrosarcomatous components; because the myoid areas were distributed throughout the tumour, the authors concluded that these cells form their own areas. O’Connell et al. studied two cases of fibrosarcoma arising in DFSP and came to a similar conclusion. Other researchers have claimed that myoid differentiation is solely a reactive phenomenon, with stromal myofibroblasts showing some degree of hyperplasia. Sanz-Trelles et al. concluded the myoid areas in DFSP to be the result of vascular smooth muscle cell hyperplasia or the proliferation of pericytes. Another study demonstrated a close relationship between blood vessels in the tumour and areas of myoid differentiation. Ohtani et al. found smooth muscle actin-expressing myoid cells in a case of fibrosarcomatous DFSP which had metastasised to the lung; the researchers suggested that myoid differentiation may therefore be neoplastic in origin. However, no relationship between the myoid areas and the blood vessels was found; furthermore, these areas differed in appearance from the pericytes.
The pigmented subtype of DFSP, also known as a Bednar tumour or storiform neurofibroma, contains varying amounts of melanotic pigment. This subtype is different from conventional DFSP and has specific chromosomal abnormalities. However, the origin of pigmented DFSP is still unknown. Some researchers advocate a neuroectodermal origin due to the presence of Schwannian differentiation along with dendritic melanocytes, while others believe that such tumours arise after trauma, scarring from vaccinations or insect and animal bites. Although several cases of DFSP with fibrosarcomatous differentiation are available in the literature, the current case appears to be the first report of recurrent DFSP with myoid differentiation and pigmentation.

Conclusion

Myoid differentiation is a morphological variant of DFSP. In the current case, myoid differentiation was found around the blood vessels in the tumour, distributed irregularly without forming a specific stromal pattern. It is possible that hyperplastic myofibroblasts occur due to a reactive process to proliferating tumour cells. However, further studies are needed to enhance our understanding of myoid differentiation and its clinical prognosis in DFSP cases.

References