

Orbital dermatofibrosarcoma protuberans with frontal and ethmoid sinus involvement: A case report and brief review of literature

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Dermatofibrosarcoma protuberans is a soft tissue sarcoma that is dermal in origin. The incidence is <0.1% of all malignancies and 1% of soft tissue sarcoma. Most commonly, it involves trunk (62%) followed by extremities (25%) and head and neck (13%). It is a slow growing tumor with locally aggressive behavior. Here, a 50-year-old female diagnosed with orbital dermatofibrosarcoma developed extra-axial component in right frontal region even on chemotherapy. Hence, the bad prognostic factors are yet to be established in dermatofibrosarcoma protuberans.

Key words: Dermatofibrosarcoma protuberans, imatinib, locally aggressive, sarcoma

Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue sarcoma which is dermal in origin, accounting for <0.1% of malignant lesions and 1% of soft tissue sarcoma.^[1] It is locally aggressive and extends into the subcutaneous tissues and muscles.^[2] It usually occurs in the second to sixth decade of life and males are more affected compared to females.^[3]

Here, we are presenting a rare case of orbital DFSP with frontal and ethmoid sinus invasion.

Case Report

A 50-year-old female presented in the radiation oncology OPD in May 2014 with complaints of swelling on the medial aspect of the right eye, slowly progressive for the last 4 months and associated with pain and diminution of vision. She also had episodes of nasal bleeding for the past 2 months. There was no other complaint of vomiting, dizziness, headache, seizures, or dyspnea. On clinical examination, approximately, 5 cm × 5 cm swelling was present over right medial canthus, displacing right eye outward with ulceration of cornea and swelling

of lower eyelid, another 2 cm × 2 cm firm swelling was also present over the medial canthus of the left eye. There was no preauricular or cervical lymphadenopathy [Fig. 1].

A direct nasal endoscopy (DNE) was suggestive of the right uncinate polypoidal polyp arising from medial turbinate.

Magnetic resonance imaging scan was suggestive of homogeneously enhancing mass lesion (5.4 cm × 3.8 cm × 4.1 cm) in right orbit with extension to ethmoid and frontal sinus, but there was no evidence of any intracranial extension lesion. Similar but small enhancing lesion was noted in the region of left medial canthus.

Histopathology was suggestive of DFSP. Spindled tumor cells are quite uniform in appearance with elongated nuclei, little or no pleomorphism with mitotic Figure and may be arranged in a storiform manner [Figs. 2 and 3]. There were characteristic positivity for CD34 [Fig. 4] and vimentin [Fig. 5] and negativity for CD31 and smooth muscle actin.

The case was discussed in the multidisciplinary clinic and was taken for palliative intent. She received palliative radiotherapy 20 Gy in 5 fractions in May 2015 and then was put on tablet imatinib 400 mg/day. Response assessment after 2 years was done, contrast-enhanced computed tomography orbit and paranasal sinuses was suggestive of progressive disease with extra-axial component in the right frontal region [Fig. 6]. In view of disease progression,



Figure 1: Swelling involving right orbit. Review of literature of orbital dermatofibrosarcoma

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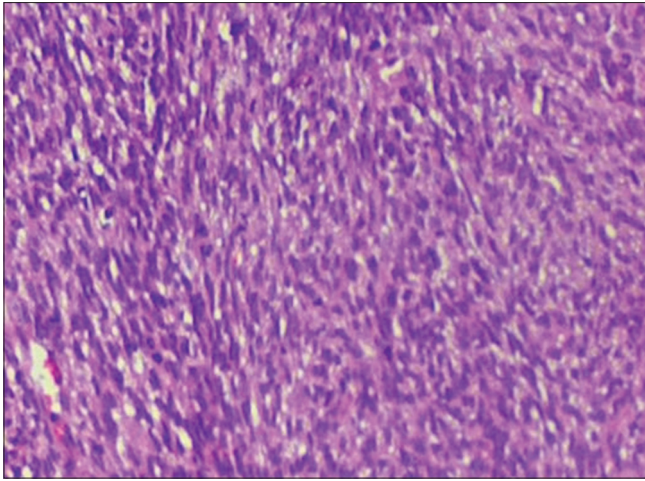


Figure 2: Histopathology of tumor showing spindle cell with nuclei arranged in storiform pattern

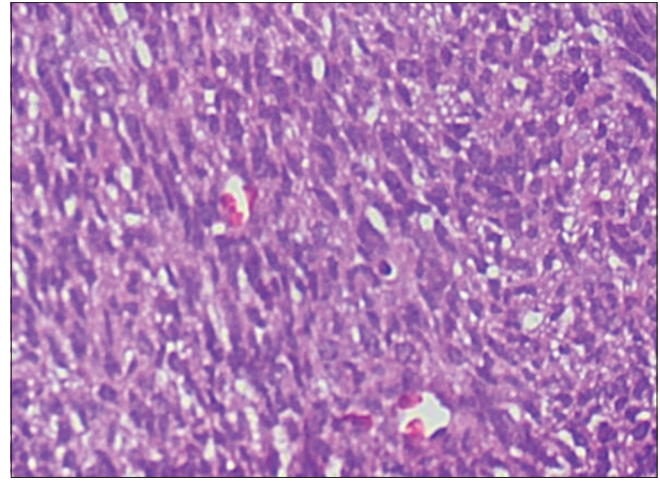


Figure 3: Histopathology of tumor showing multiple mitotic figure

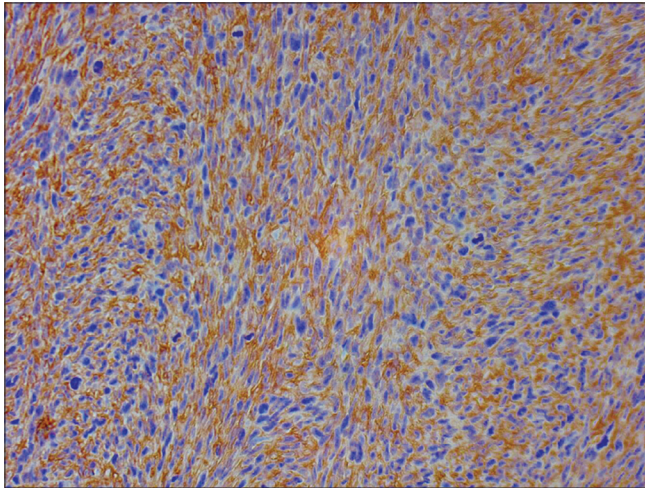


Figure 4: Immunohistochemistry staining with CD34

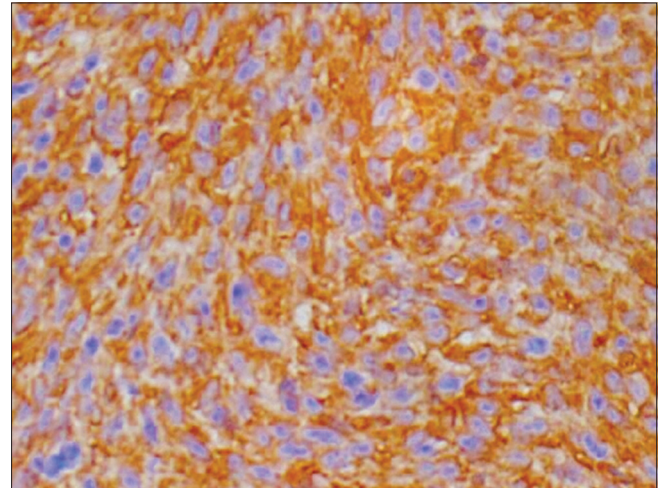


Figure 5: Immunohistochemistry staining with vimentin

the dose of imatinib has been increased to 800 mg/day. On the last follow-up, disease was stable on tablet imatinib 800 mg/day.

Discussion

The annual incidence of DFSP ranges from 0.8 to 5 cases/million population.^[4] It is dermal in origin and presents as a plaque/papule which develops into lumpy nodule over time. It usually occurs in trunk (62%), followed by extremities (25%) and head and neck region (13%).^[5] In 90% of DFSP, there is reciprocal translocation at t(17;22)(q22;q13) leading to the fusion of genes collagen Type I, alpha 1, and platelet-derived growth factor (PDGF) beta-chain resulting in activation of PDGF receptor protein tyrosine kinase. The diagnosis of DFSP is confirmed by histopathology in which proliferation of spindle cells embedded in collagen and arranged in storiform or cartwheel pattern is characteristic. DFSP is CD34 and vimentin positive.^[3]

Fibrous histiocytomas are the most common orbital tumors in adults. They develop insidiously and can be locally infiltrating, and although they are benign lesions, their rate of recurrence is high. These are characterized by a

proliferation of fusiform cells in the dermis, constituted by a variable combination of fibroblasts, collagen, histiocytes, and blood vessels. They are usually CD34 negative. Hemangiopericytomas usually appear in young and middle age and appear histologically as dense, hypercellular tumors with spindle-shaped cells. The tumors are vascular with a variably dilated, vascular branching pattern classically described as “staghorn vessels.”^[6]

The treatment of choice in DFSP is surgical excision.^[7] Mohs micrographic surgery is preferred as it preserves cosmesis.^[8] Radiation therapy is used in adjuvant setting up to a dose of 50–60 Gy in case of a positive margin or residual disease.^[9] In the present case, as the lesion was inoperable, the patient was planned for palliative radiation therapy.

Alternate treatment option for DFSP is chemotherapy. A PDGF receptor inhibitor, imatinib mesylate has been used in the past in case of locally advanced or metastatic disease.^[9,10] Other tyrosine kinase inhibitors such as sunitinib and nilotinib have been developed. DFSP of orbit is very rare. So far, only six case reports have been published in literature

Table 1: Review of literature of orbital dermatofibrosarcoma

Age/sex	Site	Surgery	Radiotherapy	Chemotherapy	Author
38/male	Medial canthus	Exentration and dacryocystectomy	-	-	Goshe <i>et al.</i> , 2012 ^[11]
70/female		Exentration	-	-	Rahman <i>et al.</i> , 2013 ^[12]
72/male					Gonnering and Sonneland 1987 ^[13]
63/male	Intracranial extension	En block excision	60 Gy	Imatinib	Bashir <i>et al.</i> , 2015 ^[14]
60/female		Excision	-	-	Brazzo and Saffra 2004 ^[15]
45/female		Surgery	-	-	Schittkowski and Wrede 2013 ^[16]
50/female			Palliative radiotherapy	Imatinib	Present study

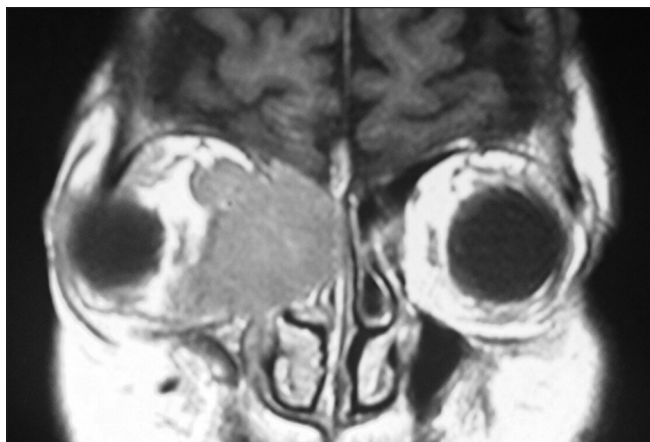


Figure 6: Contrast-enhanced computed tomography orbit and paranasal sinuses showed progressive disease with extra-axial component in the right frontal region

[Table 1]. In all six cases, surgery was the main modality of treatment except in a study by Bashir *et al.* in which adjuvant radiotherapy of 60 Gy in 30 fractions was given as adjuvant therapy followed by adjuvant chemotherapy with imatinib mesylate 800 mg/day.^[14]

Conclusion

To the best of our knowledge, this study is the first in which DFSP of orbit has been extended to involve the frontal and ethmoid sinuses. Being inoperable and locally advanced, it was decided to continue with palliative radiotherapy followed by therapy with imatinib mesylate.

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Conflicts of interest

There are no conflicts of interest.

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