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EXTRA GIANT DERMATOFIBROSARCOMA PROTUBERANS OF SCALP-CAN IT GROW BIGGER?

Neurosurgery

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ABSTRACT

Background: Dermatofibrosarcoma protuberans (DFSP) is a rare monoclonal cutaneous soft tissue sarcoma, an intermediate grade neoplasm. A massive size DFSP diagnosed in scalp region is a rare finding to come across. It typically involves the trunk and proximal extremities; rarely head and neck. It mostly occurs in second and fifth decade of life, more common in men than women. DFSP grows in a locally aggressive manner and has an increased propensity for local recurrence. The optimal treatment modality for dermatofibrosarcoma protuberans is wide excision of neoplasm. Other treatment modalities such radiotherapy and certain systemic therapies such as tyrosine kinase inhibitors can be used under special circumstances. **Case Description:** A 50-year-old male presented with a massive swelling over his scalp undergoes wide excision of tumor. **Conclusion:** DFSP in the head and neck region is characterised by a slow-growing pattern, locally aggressive in nature with high recurrence rates, and low malignant potential. This case is a unique presentation of massive DFSP of scalp.

KEYWORDS

giant DFSP, scalp, tumor, excision, CD34, recurrence

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare monoclonal cutaneous soft tissue sarcoma of mesenchymal origin , an intermediate grade neoplasm[1,2]. It mostly presents in third to fifth decade of life, but there are cases being reported from other age groups as well, including some congenital cases[3]. It's incidence is 0.1% of all cancers and 2% of all the soft tissue sarcomas, hence; it really is a rare entity to come across[4]. It typically involves the trunk and proximal extremities; rarely head and neck. Incidence rates have been reported to be nearly twice as high in black people compared to white people[5]. It is diagnosed by skin biopsy[3]. Immunohistochemically DFSP is positive for CD34 and histologically, it has characteristic morphology, of storiform islands of bland spindle cells[6]. DFSP is a result of chromosomal translocation, which leads to the fusion of protein COL1A1-PDGFB, which results in tumor growth due to overproduction of platelet-derived growth factor (PDGF)[3]. Cytogenetic analysis of some DFSP tumors shows t(17;22) of ring chromosome[7]. DFSP grows in a locally aggressive manner and it has an increased propensity for local recurrence but it rarely metastasize[3]. The optimal treatment modality for dermato fibrosarcoma protuberans is wide excision of neoplasm.[3].

As it is a neoplasm of high recurrence rate, wide excision is the key to reduce its recurrence[7]. In cases of unresectable, recurrent, or metastatic dermatofibrosarcoma protuberans, imatinib mesylate a chemotherapeutic agent is currently FDA-approved for adults[8]. Adjuvant radiotherapy (RT) has been suggested to reduce the risk of local recurrence in neoplasm with narrow or positive surgical margins[9].

Case Report

A 50 year old male presented to a tertiary care hospital in New Delhi, India; he presented with a swelling over his scalp since 6 months; it was initially small in size which subsequently increased in size within six months. He described it to be painful accompanied with pressure sensation; few ulcerated lesions were present over the swelling, and it was associated with headaches. He did not had any history of trauma and due to the massive size of swelling at back of his head it was difficult for him to sleep in supine position.

Figure 1 Swelling in the left occipital region in a 50 year old male patient

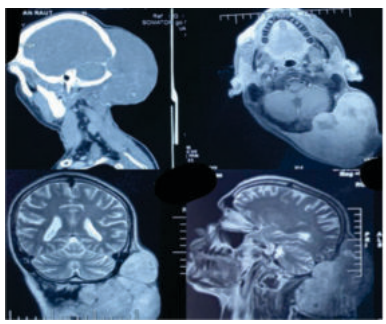

Figure 2 Contrast enhanced CT scan of head.

On examination, his scalp swelling was found to be enormous in size, it measured about 22cm, with extensive ulcerations. Overlying skin showed necrotic areas with crusting, exudative secretions and areas of haemorrhage(Figure 1). External surface of mass was nodular. It was a cosmetically disfiguring swelling. He was subsequently operated under general anaesthesia, and was intubated in lateral position; it was a challenging case for anaesthesia team as well. He successfully underwent wide radical local excision with removal of involved occipital bone and local flap reconstruction with excellent cosmetic outcome (Figure 3). The tumor was highly vascular which resulted in

massive blood loss, he was successfully managed with IV fluids and blood transfusion. Histopathological report suggested, tumor cells to be arranged in intersecting fascicles and vague storiform architecture; they were spindle shaped showing palisading arrangement having eosinophilic fibrillary cytoplasm with areas of necrosis and few mitotic figures. Immunohistochemically cells were positive for CD34.He is currently being followed up with neurosurgery for any recurrence and is undergoing radiotherapy.

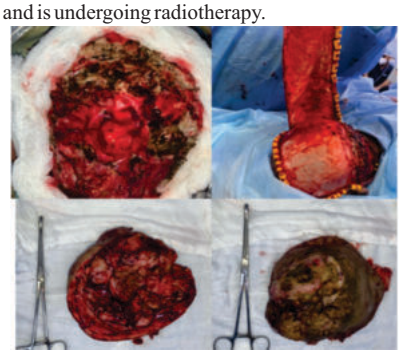

Figure 3 Intraoperative finding showing soft tissue neoplasm. Wide radical local excision of tumor. DFSP size approximately 22x14x10 cm.

DISCUSSION

DFSP is a slow growing tumor, locally aggressive, intermediate grade soft tissue neoplasm with a pronounced tendency for local recurrence, that rarely metastasizes[10]. Excess of PDGF-β is considered to play a role in the development of dermatofibrosarcoma[11]. It is most often diagnosed in individuals aged between 30 to 50 years ,however there are few cases being reported from other age groups too[3,12]. DFSP arises in a healthy skin or in areas exposed to repeated trauma, radiations or scarring [13]. It most commonly involve the trunk and proximal extremities whereas the head and neck region is less commonly involved; and is more common in men than in women [1,13]. It is a locally aggressive tumor with high risk of local recurrence[14]. As a result it has direct implications for surgical management, which is the gold standard for DFSP directly affecting the prognosis of disease[3,15]. Patients presenting with scalp DFSP, the differential diagnosis could be midline embryologic defects, such as glioma, dermoid cyst and cephalocele[10]. Immunohistochemically CD34 is an important immunohistochemical marker for diagnosing DFSP[16]. Cytogenic analysis shows t(17;22) in some of these tumors[7]. The recurrence potential of DFSP is related to the extent of resection, but it can be very difficult to determine a positive margin because DFSP has fibroblast-like morphology[12]. However, tumors; that are removed by wide local excision with clear margins represent a reduced subset of lesions, as seen in high grade of sarcomas[15]. There is higher risk of subclinical extension and local recurrence with enlarged tumor size and depth [17]. DFSP's preferred treatment modality is surgery, however there are still chances of local recurrences which warrants the need for lifelong surveillance[14].Other treatment modalities such as, Radiotherapy and other systemic therapies like tyrosine kinase inhibitors can be used under special circumstances[7] .

Figure 3 Postoperative scar tissue.

CONCLUSION

DFSP of scalp with this massive size is a rare finding and difficult to diagnose; especially when it involves head or neck which is a rare site for it to occur. As a result, its treatment is often delayed because of misdiagnosis. Therefore, one must be aware of this rare condition and perform wide excision for these tumors to reduce the recurrence rate.

A giant scalp DFSP is rare and difficult to diagnose, particularly when presenting at the head and neck region. Treatment of giant scalp DFSP is often delayed because of misdiagnosis, leading to wide local excision resulting in a significant functional or cosmetic deficit. Surgeons should be prepared for reconstruction following tumor removal. Hence, we should be aware of this uncommon entity, and always perform a wide excision of these neoplasm to reduce the risk of recurrence. It should be diagnosed and treated at an early stage for a better outcome. Hence, the diagnosis of DFSP should always be considered in cases of massive scalp swelling.

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