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Dermatofibrosarcoma protuberans: A rare presentation in the scalp. Case report

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SUMMARY

Background. Dermatofibrosarcoma protuberans of the scalp (DFSP) is a rare soft tissue neoplasm originating from the dermal layer of the skin, usually affecting the adults.

Case report. The current case report presents a 48-year old male with a huge lump on the right side of parietal region. A wide local excision of the tumor was performed and the excised specimen was sent for histopathological examination. Histopathology and Immunohistochemistry was suggestive of DFSP.

Conclusion. Dermatofibrosarcoma protuberans is a rare neoplasm affecting the head and neck region. This unusual entity is more likely to recur when a small margin of surgical excision is performed. Wide local excision is the gold standard treatment and radiotherapy is preferred in recurrent diseases.

Keywords: scalp, wide excision, storiform pattern, CD4, vimentin.

INTRODUCTION

DFSP is an unusual neoplasm most often diagnosed in individuals between 20 to 50 years of age with trunk (50%-60%) and proximal extremities (20%-30%) are the most common site of occurrence (1) and less commonly presenting with head and neck region (2-4). DFSP has an aggressive growth pattern with a high propensity for recurrence (20%-50%) after surgical excision. Histologically, the tumor features a storiform growth pattern with uniform tumor cells, hyperchromatic areas, enlarged nucleoli showing a classic honeycomb appearance infiltrating into the subcutaneous tissue. Immunohistochemical studies show positivity for CD4 and Vimentin. Several reports in the literature suggest that wide local surgical excision of 3-5 cm surgical margins would ensure good prognosis and reduce the risk of recurrence (2).

CASE REPORT

A 48 year old male presented with a painless swelling in his right parietal region for the past one year. On clinical examination, a single, non-tender, firm nodule measuring 5 cm in diameter, mobile on palpation with base fixed. The swelling was non-pulsatile and trans-illumination was negative. Magnetic resonance imaging showed a soft tissue mass in the right parietal region without any extension into the cranium (Fig. 1). A wide local surgical excision was performed incising the skin, subcutaneous tissue to the underlying periosteum ensuring 3cm clear surgical margins. The defect was then undermined with a blunt tipped scissors to minimize the tension on the wound margins. The whole specimen was sent for histopathological investigation (Fig. 2). Hematoxylin and eosin stained sections showed a dense cellular and poorly defined tumor in the dermis comprising of uniform spindle shaped cells, oval nuclei, vesicular chromatin and moderate cytoplasm with classic feature of cartwheel appearance (Fig. 3). Immuno-histochemical studies showed tumor cells positive for CD4 and vimentin (Fig. 4) respectively.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) was first named by Hoffmann (5). The tumor is otherwise represented as hypertrophic morphea, sarcomatous tumor as it resembles a keloid, progressive and recurrent dermatofibroma (6). It mostly occurs in the 2nd and 3rd decade, affecting both the gender groups. Burkhardt *et al.* (7) reported 6 out of 56 cases involving the scalp and Tamoney *et al.* (8) found 40 out 261 cases affecting the head and neck region. While, Mbonde *et al.* (9) reported with higher incidence of cases, particularly affecting the scalp implicating trauma as the primary cause.

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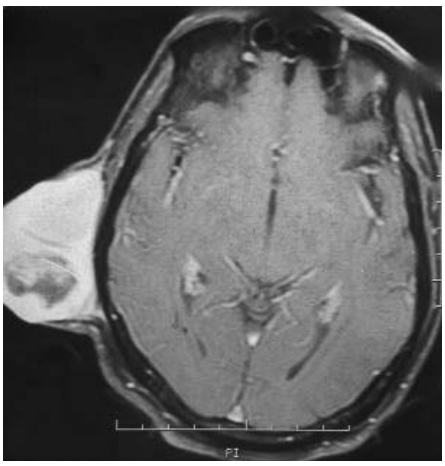

Fig. 1. Magnetic resonance imaging showing a soft tissue density mass in the right parietal region

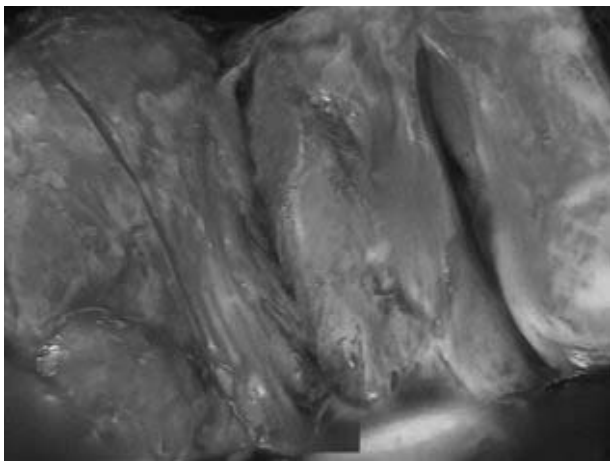

Fig. 2. Excised soft tissue mass from the right parietal region

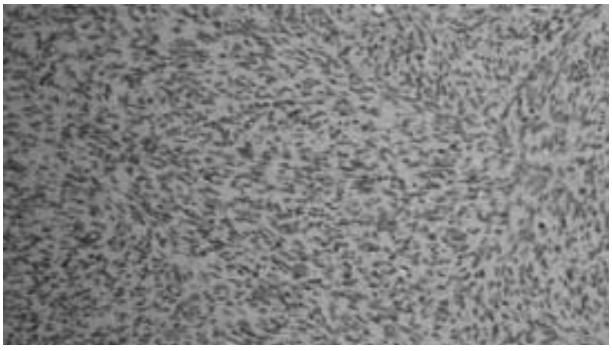

Fig. 3. Haematoxylin and eosin stained section showing typical storiform pattern of dermatofibrosarcoma protuberans

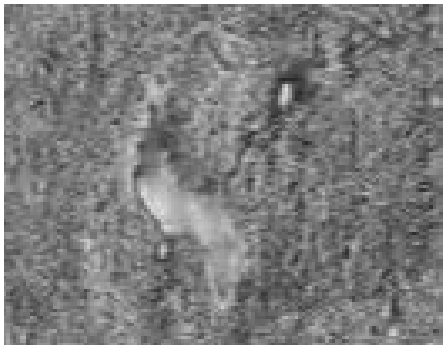

Fig. 4. Immunohistochemistry section showing tumour cells positive for CD 34

DFSP usually presents as an asymptomatic swelling with a history pertaining to slow persistent growth. The swelling gradually increases into a lumpy nodule and over time it develops into an ulcerative protuberant tumor. During the initial stages of growth the tumor is mobile on palpation as it is adhered to the overlying skin and not to the deeper structures. At the latter stages, the tumor progresses infiltration into the fascia and muscle indicating a base fixed tumor. Telengiactasis may also be present at the periphery. The tumor was misdiagnosed at the time of presentation as it may be without symptoms or a state of confusion with other benign dermal lesions. The differential diagnosis includes lipomas, epidermal cysts, keloid, dermatofibroma and nodular fasciitis at the initial stage of tumor growth and at late stages when it becomes protuberant, it should be differentiated from pyogenic granuloma, Kaposi sarcoma, schwannoma, neurofibroma and other soft tissue sarcomas (10, 11).

Histologically, the tumor is cellular and comprised of monotonous spindle shaped cells with elongated nucleoli showing few or absence of cellular pleomorphism or nuclear hyperchromatism. The cells are arranged in an irregular fashion with a characteristic honeycomb pattern. Uncommon variants of DFSP are myxoid features of DFSP and pigmented Bednar tumor with the presence of melanin containing dendritic cells dispersed throughout the tumor (12, 13). Immunohistochemistry demonstrates DFSP positivity for CD 34 and negativity for S-100 proteins (13-15). The fibrosarcomatous areas characterize a herring bone growth pattern rather than a storiform pattern which possess a foci of hypercellularity, increase in cell size and mitotic activity. Ugurel *et al.* (16) proposed a staging for DFSP as per the German guidelines. Stage I represents a primary stage tumour, stage II describes a regional lymph node metastases and stage III and IV characterize a distant metastasis (11).

The treatment of DFSP is wide local surgical excision with negative margins. Wide excision 2-4 cm of clinically uninvolved skin, including fascia must be achieved in order to minimize the local rate of recurrence (17). Mohs micrographic surgery (13, 18) is routinely practiced which allows precise mapping of the surgical margins both in depth and lateral histologically. This technique would involve continuous sequential sections of the resected tissues being subjected for frozen microscopic examination until free surgical margin is obtained. At this stage, primary closure is not advised hence reconstructive surgeries are performed using skin graft, local skin flap and myocutaneous flap. Neck dissection is not preferred unless a regional lymphadenopathy is suspected. However, a sentinel lymph node biopsy would be recommended in DFSP-FS (fibrosarcomatous) (14).

DFSP is non-sensitive to chemotherapy and radiotherapy may be a treatment of choice for unresectable lesions. Rutkotvski *et al.* (19) reported a partial

response for up to 50% using Imatinib and Ng et al (20) using vinblastine and oral methotrexate once a week for more than six weeks and a follow up for 5 months. Sheidaei *et al.* (15), stated that in addition to Imatinib, Sorafenib would be successful for patients with DFSP, who are unresponsive to radiotherapy postoperatively. Minter *et al.* (21) have reported a high recurrence rate in tumor margins <2.5 cm and reports on distant metastases to bone, lungs and sometimes pancreas have also been witnessed in English literature (22). Suryawisesa *et al.* (17), emphasized that tumor with positive surgical margins showed a higher recurrence rate of 20% to 50% while, Wiesmueller *et al.* (13) encountered no recurrence or metastasis over a follow up of 84 months.

CONCLUSION

Dermatofibrosarcoma protuberans is a rare neoplasm affecting the head and neck region. This unusual entity is more likely to recur when a small margin of surgical excision is performed. Wide local excision is the gold standard treatment and radiotherapy is preferred in recurrent diseases.

COMPLIANCE WITH ETHICAL STANDARDS

Conflict of interest

The authors declare that they have no conflict of interest.

Declaration of patient consent

The authors certify that they have obtained written informed patient consent for the surgery under general anesthesia and has given his consent for images and other clinical information to be reported in the journal with an understanding that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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