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RESEARCH ARTICLE

MYXOAID DERMATOFIBROSARCOMA PROTUBERANS: A RARE DIAGNOSIS AND REVIEW OF LITRATURE

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ABSTRACT

Dermatofibrosarcoma protuberans is a very rare slow growing deceptive neoplasm. It is reported that the classic form of dermatofibrosarcoma has a very limited metastatic potential, while a fibrosarcomous transformation of dermatofibrosarcoma protuberans has a much higher potential. Dermatofibrosarcoma Protuberans (DFSP) represents a low grade, locally aggressive mesenchymal neoplasm with characteristic clinicopathologic, immunohistochemical, and molecular findings. Myxoid DFSP is an uncommon variant, with only few cases reported in the literature and may present a diagnostic challenge on histopathologic examination. It has potential to develop local recurrence as well as metastases especially to the lungs. A 27-year-old man presented with soft tissue swelling on right of temporal region for one month. It was 3X2 cm. of size and firm on consistency. The swelling was mobile on underlying structures and was painless. The provisional diagnosis of sebaceous cyst was made and excision biopsy done with full margins of overlying skin and deep up to muscle. We report here the case of myxoid DFSP along with a review of literature arising in the scalp describing the morphologic and immunohistochemical findings. Recognition of this variant at unusual sites is clinically important because the differential diagnosis includes benign and malignant tumors which could lead to under or over-treatment.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare slow growing neoplasm of the skin. Darier and Ferrand first described this tumour in 1924 as a distinct entity and named it as progressive and recurring dermatofibroma. Hoffman was responsible for naming the tumour as dermatofibrosarcoma protuberans in 1925 [1]. It accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas [2]. Dermatofibrosarcoma Protruberans (DFSP) is a superficial, low grade locally aggressive mesenchymal neoplasm of fibroblastic origin which arises in the dermis and frequently involves the subcutaneous tissues. It arises most commonly in the trunk, extremities, and head and neck but may involve any body site [3]. Several histologic variants are recognized including pigmented DFSP (Bednar tumor), fibrosarcomatous DFSP, myxoid DFSP, flat atrophic DFSP, giant cell fibroblastoma, and DFSP with myogenic differentiation [4]. The Myxoid variant of DFSP represents a rare morphologic variant with prominent myxoid stromal changes that may cause considerable diagnostic challenges, particularly in the distinction from other more clinically aggressive myxoid mesenchymal neoplasms.

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To the best of our knowledge, this is the first case of myxoid DFSP involving the scalp. We describe the morphologic and immunohistochemical findings and discuss the differential diagnosis of this rare variant of DFSP.

Case Report

A 27-year-old man presented in our outdoor with swelling on right side of temporal region for six months. On examination, it was 3X2 cm. of size and soft to firm in consistency. The swelling was mobile on underlying structures but adherent to overlying skin and was painless. The provisional diagnosis of sebaceous cyst was made and excision biopsy done with full margins of overlying skin because skin was adherent to swelling and it was deep to subcutaneous tissue but not involving the muscle. Excised mass was look like fibro-fatty tissue and no cyst wall or fluid was found. The specimen was sent for Histopathological examination. Microscopic examination show a tumor composed of oval to spindle cells arranged in sheets, fascicles and stained form pattern against a myxomatous stroma and contain hyaline, blood vessels and plexiform capillary network. Tumor is mitotically inactive and at places infiltrated into surrodung adipose tissue. On immunohistochemical examination- tumor cells are- Vimentin-positive, CD 34-positive, SMA-positive in blood vessels, S: 100-negative, CD 63-negative, Desmin-negative. These features are suggestive of Dermatofibrosarcoma protuberance. All the sutures were removed on seventh post operative day and Post-operative period was uneventful. (figures-1&2 showing complete healing of wound) Histopathological

examination of the tumor revealed myxoid dermatofibrosarcoma protuberans with clear resection margins.

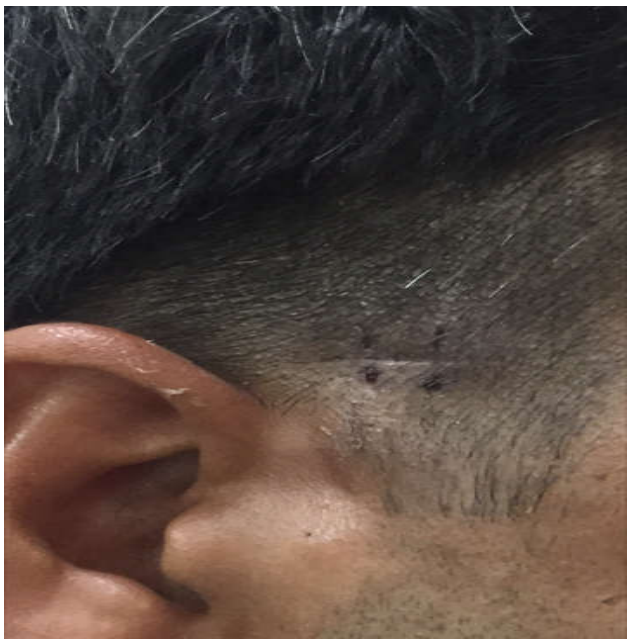

Figure 1 Showing Post Excision After Stich Removal

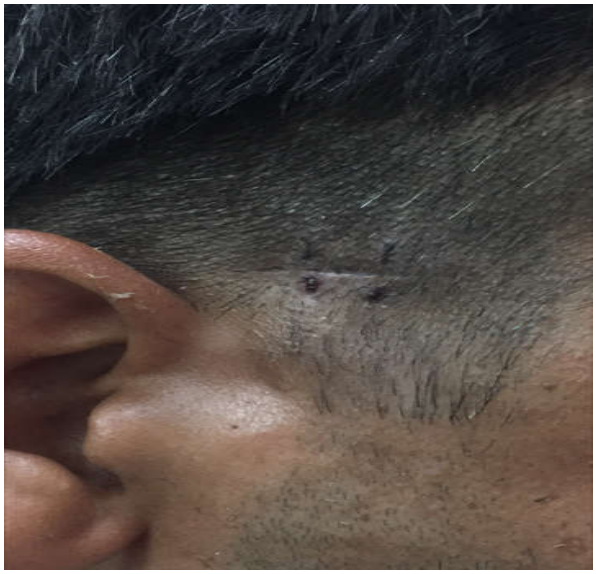

Figure 2 Complete Wound Healing

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a low to intermediate – grade cutaneous sarcoma which is rare in adults [5].In the WHO classification of the mesenchymal tumors, DFSP is an intermediate-grade, locally aggressive tumor which recurs commonly but metastasizes very rarely [6]. The annual incidence of DFSP as reported in literature based on population based cancer registries of various countries is less than three cases per million populations making it a rare tumor [7]. The Myxoid variant of DFSP was first described by Freirson and Cooper in 1983 [8] who reported two cases of myxoid DFSP that showed distinctive histologic features including a monotonous haphazard arrangement of spindle-shaped and stellate tumor cells with little variation in cellularity from field to field, pale basophilic stroma containing hyaluronic acid, minimal cytologic pleomorphism and a low mitotic rate with a dispersed, non-patterned vasculature.

Distinctive histologic features of myxoid DFSP, present in our case include myxoid stroma and prominent thin-walled vessels, alternating with relatively hypercellular areas forming a storiform pattern. Cells were embedded in the myxoid stroma in a haphazard pattern and had a spindle appearance with indistinct nucleoli.

Reimann and Fletcher [9] defined myxoid DFSP as DFSP with greater than 50% myxoid stroma. Immunohistochemically, in addition to the characteristic and diagnostic CD34 expression, expression of SMA was noted. These findings are consistent with the immunohistochemical profile of typical DFSP [10]. Tumor cells were negative for cytokeratin, ALK-1, desmin, S100, CD31 and MUC4 immunostains which was helpful in supporting the diagnosis of myxoid DFSP. The histologic differential diagnosis of myxoid DFSP is diverse and includes benign and malignant tumors. Among the benign tumors, superficial angiomyxoma, myxoid neurofibroma, superficial acral fibromyxoma, nodular fasciitis and solitary fibrous tumor should be considered. Superficial angiomyxoma can be difficult to distinguish from myxoid DFSP, particularly on a small biopsy, as both tumors have a myxoid stroma rich in blood vessels and express CD34 on tumor cells. However, typical cases of superficial angiomyxoma tend to be less cellular, have a lobular rather than the infiltrative growth pattern seen in myxoid DFSP and contain neutrophils as well as epithelial structures in 25% of the cases [11]. The variants described in the literature include the atypical, palisading, keloidal, granular cell, myxoid, lichenoid, balloon cell and signetring cell variants [12].

Frierson and Cooper [13] Reported the first case of the rare DFSP myxoid variant in 1983, and Hong *et al*. [14] showed another case of this clinical form rarely documented in the literature, with prominent myxoid stroma alterations. Tantcheva-Poor *et al*. [15] described a vascular histological variant of DFSP. Martin *et al* reported that in almost 50 % of their patients the tumor presented at first as a "non-protuberant" DFSP, with a mean period of 7.6 (± 9.3) years before developing into a protuberant DFSP [16]. The proportion of involvement between men and women is 1:1; however, Asquo *et al*. [17] reported a slight predominance in the male gender the primary treatment for DFSP is complete surgical resection having a very low recurrence rate of 1.5%. The best method to remove these lesions is the Mohs micrographic surgery [18]. We reported here a case of DFSP along with a review of literature. In our case, there is no infiltration of the adjacent structures like muscle or bone was seen. On ultrasound, DFSPs have been found to be mostly hypo echoic or mixed hyper echoic, with mostly well-defined margins or irregular, with projections similar to pseudopodia [19]. This happened in our case, where a preoperative diagnosis of lipoma was made. MRI studies are also not specific since they may not always distinguish DFSPs from other soft tissue sarcomas [20]. Therefore, histological examination is the only definitive diagnostic method. Microscopically, DFSP is characterized by diffuse infiltration of the dermis and sub cutis, usually sparing the epidermis and skin appendages. It grows along preexisting fibrous septa while infiltrating fat lobules giving a typical honeycomb pattern. Rarely DFSP might present as an infiltrative subcutaneous mass [21].

Diagnosis of this tumor can be done by FNAC. But exact details are best revealed by histopathological examination of the specimen. Metastatic workup in cases of recurrent tumors

involves evaluation of lungs followed by lymph nodes. Magnetic resonance is the investigation of choice for evaluating the extent of local spread [22]. However, lymph node involvement is extremely rare. Surgery is the treatment of choice for fresh cases. The size of the tumor dictates the extent of resection and the need for reconstruction thereafter [23].

Myxoid DFSP tends to have a good prognosis with low metastatic potential; however, they are locally aggressive with high rate of local recurrence following incomplete excision. [24]. In our case, tumor cells tested positive for vimentin and CD34, thereby setting the diagnosis of DFSP. In cases with possible bone involvement, the periosteum or even a portion of the bone may also need to be excised to achieve negative resection margins [25]. Most local recurrences appear within the first three years of excision, with 50 % presenting within the first year of surgery. However, recurrences are also reported even after five years. Thus, it is important to follow-up these patients for long-term [26].

CONCLUSION

We report a case of myxoid DFSP a rare variant of DFPS. Because of its myxoid appearance, it may present a diagnostic challenge. Recognition of this variant is clinically important because the differential diagnosis includes both benign and malignant tumors. Sometimes diagnosis is missed and investigations are not supporting. Therefore, every swelling present over any part of body must be sent for histopathological examination after complete resection. Because of its rarity, information is lacking regarding the optimal therapy and potential utility of immunohistochemistry in diagnosis, so further investigation is needed.

Conflict of interests; *None* of the authors have declared any conflict of interests.

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