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Comprehensive Analysis of Dermatofibrosarcoma Protuberans: Case Report and Management Strategies

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Abstract: ***Dermatofibrosarcoma Protuberans DFSP is a rare, locally aggressive, soft tissue sarcoma of the skin with a low to intermediate malignancy rate. This article presents a case report of a 40yearold female with a DFSP lesion on her left breast, progressing over six months. Diagnosis was confirmed through cytology, histopathology, and immunohistochemistry, highlighting spindle cell proliferation and positive CD34 and Vimentin markers. The tumors clinical features, diagnosis methods, and treatment strategies, including surgical excision and adjuvant therapy, are discussed. Early detection and complete resection are vital for reducing recurrence rates and improving patient outcomes.***

Keywords: Dermatofibrosarcoma Protuberans, DFSP, spindle cell sarcoma, CD34, surgical excision

1. Introduction

Dermatofibrosarcoma protuberans (DFSP) is the most common sarcoma of the skin. DFSP is a relatively uncommon soft tissue neoplasm with low to intermediate grade malignancy. Although distant metastases are infrequent, DFSP is highly aggressive locally with frequent local recurrences. It constitutes less than 0.1% of all cutaneous malignancies and 1% of all soft tissue sarcomas. DFSP is commoner in males than in females with a peak incidence during the third decade of life. The most common body part affected is trunk followed by upper limbs and head and neck. It may also occur at sites of previous trauma. The main chromosome involved in its pathogenesis are Chromosomes 17 and 22. The basic pathophysiology is Translocation t (17: 22) involving COL1A1 (collagen type 1α 1 gene) and platelet-derived growth factor β (PDGF β) genes. DFSP is a clinically cutaneous fibrous tumor that exhibits a slow and infiltrative pattern of growth. It has a marked tendency to recur locally after surgical excision. Some of the rare cases can even metastasize to regional lymph nodes or distant sites. Local recurrences occur in 20-55% of cases. Early diagnosis, wide excision and regular follow up are essential components of management of DFSP.

2. Case Report

A 40 year old female presented with a lesion over left breast. The swelling was pea sized but progressed gradually over a period of 6 months. On examination the lump was revealed to be firm, immobile and 3 cm in diameter. The overlying skin was reddish in colour. The radiography was non specific. MRI stated an isointense hypo echoic lesion with an unknown site of origin. Fine needle aspiration of the lesion was done and the smears were prepared. The smears were stained with leishman giemsa stain. On examination the cytology slides showed presence of abundant spindle cells in streaming pattern at places. Post FNAC, a small biopsy of the lump was done for definite diagnosis. The slide was stained with haematoxylin and eosin stain. The section showed stratified squamous epithelial lining. The subepithelium showed proliferation of long oval to spindle cells with long slender nuclei and moderate amount of eosinophilic cytoplasm. The adjacent stroma was fibrocollagenous with inflammatory cells and multiple blood vessels. On further work up Immunohistochemistry was done. CD 34 and Vimentin markers were applied. Both were strong positive. Hence it was concluded to be a case of Dermatofibrosarcoma protuberans.

3. Discussion

DFSP is a relatively uncommon spindle cell sarcoma of fibroblastic or myofibroblastic differentiation. It was first described by Taylor in 1890, but Hoffman coined the current nomenclature when he reported three cases in 1925. Other terms used to describe the neoplasm are: Progressive and recurrent dermatofibroma (Darier and Ferrand), hypertrophic morphea (Sherwell), sarcomatous tumors resembling keloid (Taylor), fibrosarcoma of the skin (Stout). Most of these tumors are focal in origin. The differential diagnosis for DFSP lesions are dermatofibroma, schwannoma, cutaneous neurofibroma, solitary fibrous tumor, intradermal spindle cell lipoma and spindle cell or desmoplastic melanoma.

In the early stages of DFSP, they are non - protuberant lesions but later on they turn into indurated, purple, and violaceous nodules. If left unresected, the tumor can reach the underlying tissue such as fascia, muscle, periosteum, and bone. It can even cause distant metastatic lesions in organs such as the lungs, brain, and lymph nodes. However, this process can range from a few months to decades.

Histopathologically, DFSP shows a proliferation of homogeneous spindle cells with scant cytoplasm and elongated hyperchromatic nuclei involving the dermis and subcutaneous tissue. These cells are not pleomorphic and have low mitotic activity. They are typically arranged in well - defined bands which interweave or radiate like the spokes of a wheel, forming an irregularly whorled or storiform pattern. In early stages, a grenz zone can be observed between the tumour and the epidermis. Tumour cells invade the subcutaneous tissue with irregular tentacle - like projections through septa and fat lobules and hence local recurrences after excision are common. DFSP usually stains positively for CD34 and negatively for factor XIIIa. DFSP lesions shows various forms. The atrophic and pigmented forms are rare variants. The pigmented DFSP lesions, also called Bednar tumors are dark blue

or black coloured in appearance. Such lesions have melanin containing dendritic cells besides other histological findings of DFSP. On the other hand, the atrophic variant is mostly a flat plaque rather than the usual protuberant lesions. It resembles a dermis - based skin mass with typical plaque - like histological features besides dermal layer thinning. However, these two variants have spindle cells with positive vimentin and CD34 staining. DFSP cases are usually an unexpected pathological result of a presumably benign skin mass. Such conditions can result in several years delay in diagnosis of patients with cases suspicious for DFSP. IHC CD 34 has a sensitivity of 85% in diagnosis of DFSP. Hence it has a key role in differentiating DFSP from other benign soft tissue tumors.

In terms of treatment of DFSP, complete surgical excision is the gold standard. Neoadjuvant therapy with imatinib and radiotherapy is suggested in the majority of cases that experienced incomplete resection. Such cases with incomplete resection suffer from recurrence very often. Indeed, surgical resection margins have been found to have a direct impact on the risk of future recurrence. The most crucial prognostic factor is the extent of surgical excision, which indicates the importance of extent of removal of deeper involved tissue such as deep fascia and muscles. The recurrence rate varies from 10 to 80% in various studies. However, one study reported that the recurrence was double in lesions with margins less than 2 cm in comparison with those with a larger diameter.

4. Conclusion

Dermatofibrosarcoma Protuberans DFSP remains a challenging diagnosis due to its slow progression and tendency for local recurrence. Early recognition, accurate diagnosis through histopathology and immunohistochemistry, and complete surgical excision are critical to managing this rare tumor. While DFSP rarely metastasizes, recurrence rates highlight the importance of thorough surgical removal, especially in cases with incomplete resection. Continued research on adjuvant therapies like imatinib may provide additional strategies to improve patient prognosis and reduce recurrence rates. Regular followup is essential to monitor for any signs of recurrence.

References

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