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Case Report

Anterior abdominal wall spindle cell tumour - atypical fibrous histiocytoma: a rare presentation

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ABSTRACT

Atypical fibrous histiocytoma is a rare and a distinct variant of cutaneous fibrous histiocytoma which can be misdiagnosed as sarcoma. It is mainly composed of a mixture of fibroblastic and histiocytic cells, especially found in the skin (dermatofibroma), particularly in the limbs. It is quite uncommon and is difficult to distinguish from a malignant lesion. Due to the lack of clear cut predictive morphological patterns and due to the suspicion of malignancy, complete surgical excision is recommended. Provided that atypical fibrous histiocytoma is treated by complete excision, a benign outcome is to be expected in most cases. However, like the cellular and aneurysmal variants of fibrous histiocytoma, atypical fibrous histiocytoma shows a higher tendency to recur locally than ordinary fibrous histiocytoma and may rarely metastasize. Lesions with floridly atypical features represent potential pitfalls for overinterpretation as pleomorphic sarcoma, which would appear to be inappropriate in most cases. Due to its rarity and uncertainty, we report a case of atypical fibrous histiocytoma and discuss its presentation, nature, types and treatment with reference to a brief review of literature.

Keywords: Abdomen, Fibrous histiocytoma, Malignant, Sarcoma, Tumour

INTRODUCTION

Atypical fibrous histiocytoma is a slow growing nodular polypoid neoplasm found exclusively in dermis and subcutaneous tissue.[1] It is a locally aggressive soft tissue tumour with high recurrence rates and low to intermediate malignant potential.[2] The annual incidence is 0.8 to 4.5 per million.[3] It represents 1% of soft tissue sarcomas and less than 5% of all soft tissue tumour.[4] Clinically and histologically, it is very similar to typical dermatofibroma (fibrous histiocytoma), but it is important to differentiate it from them because of the possibility of presenting local recurrences and, in some cases, development of distant metastases. This lesion is not well known and merits wider recognition in order to avoid inappropriate treatment.[5] The criteria for differentiation concern mainly the architectural pattern of the lesion rather than its cytological features.[6]

CASE REPORT

This was a 69-year-old woman who presented with a five-year history of swelling over the abdomen which gradually increased in size. She was asymptomatic with no complaints of any pressure symptoms, pain and bowel/bladder disturbances. She presented to the hospital because of a one-month history of ulceration and purulent discharge from the swelling.

Clinical examination revealed a 5 X 6 cm swelling in the midline just below the umbilicus with well-defined borders, firm in consistency, mobile, non-tender with

ulceration at the centre of the swelling with foul smelling discharge.

The plane of the swelling was above the parietal wall with mobility and no attachments to the deeper structures. However, it was fixed to the skin with ulceration of the skin. Figure 1

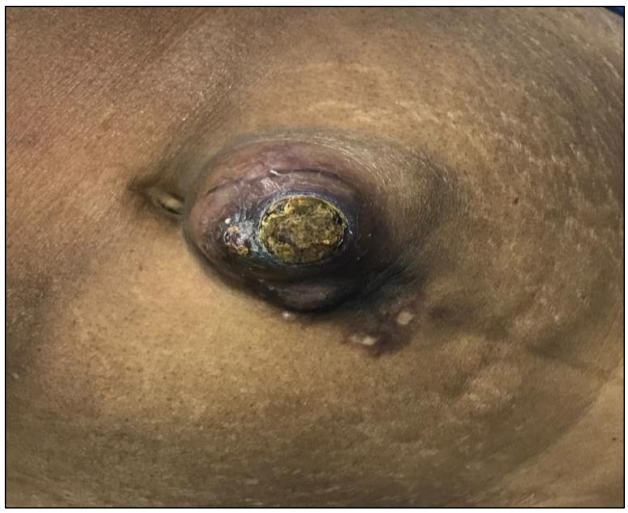

Figure 1: Ulcerated swelling fixed to the skin above the parietal wall below umbilicus.

Soft tissue sonogram reported a large well-defined hypo echoic mass lesion measuring 6.2 x 3.4 x 4.6 cm in subcutaneous plane with good vascularity. Routine blood investigations were unremarkable.

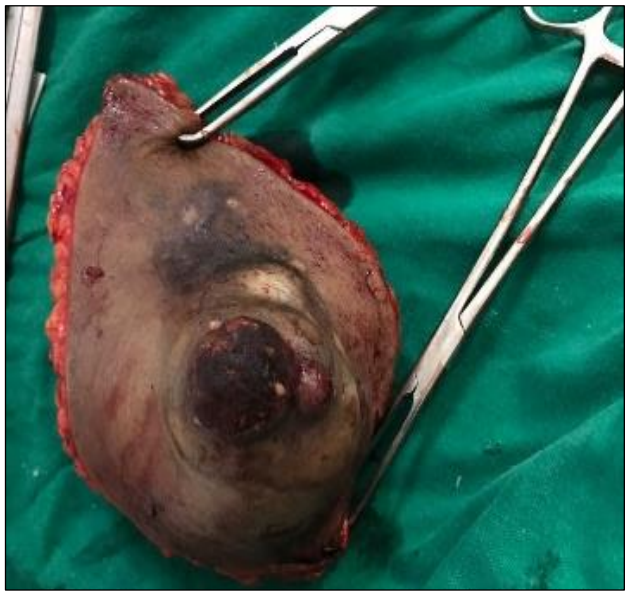

Figure 2: Specimen- post wide local excision with 2 cm margin clearance arising from subcutaneous tissue involving the skin.

The patient underwent wide local excision of the swelling with a 2 cm margin clearance with primary closure of the wound. Intra-operative finding was a vascular tumour of size 5x6 cm arising from the subcutaneous tissue involving skin (Figure 2). Post-operative recovery was uneventful.

Figure 3: Gross pathological specimen suggestive of intermediate grade spindle cell tumour: atypical fibrous histiocytoma with margins free of tumour.

Histopathology report gave an impression of an intermediate grade spindle cell tumour suggestive of Atypical fibrous histiocytoma with margins free of tumour. Figure 3

DISCUSSION

Atypical fibrous histiocytoma is a rare variant of cutaneous fibrous histiocytoma also called pseudo-sarcomatous fibrous histiocytoma or dermatofibroma with monster cells.[7,8] This tumor is seen as a solitary firm cutaneous nodule between 5 to 79 years. Anatomical distribution is wide with most cases occurring in the lower and upper extremities (79%). It is relatively mono-morphous, mononuclear, spindle cell lesion involving both dermis and subcutis. Several tumors enter the differential diagnosis including atypical fibroxanthoma, dermal leiomyosarcoma, sarcomatoid carcinoma, nodular melanoma, dermatofibrosarcoma protuberans, angiosarcoma, and pleomorphic fibroma.[9]

It grows in an infiltrative pattern, spreading along connective tissue septa and with unpredictive radial extensions often associated with satellite lesions. It has a high rate of recurrence that can occur in 20-55% of cases and with an extremely low rate of distant metastasis. Principal treatment for this is wide excision with margin of 2 cm for tumors less than 5 cm. For tumors more than 5 cm, wide excision with adjuvant radiotherapy is preferred to limit local recurrence. With aggressive

resection and with radial margins recurrence rate is about 5%.

Atypical fibrous histiocytoma is a heterogeneous process, in which histiocytoid cells probably represent the neoplastic component and fibroblastic cells may represent a reactive proliferation or alternatively, it may represent a true neoplasm in which neoplastic cell type has been obscured by prominent reactive fibroblastic component.[10]

Infiltration of superficial subcutis is not uncommon and does not affect prognosis. Clinical behavior is benign, but some variants (cellular, aneurysmal, and atypical) are associated with non-destructive local recurrence in 20% of cases.

Six of 53 lesions (11%) in the series of Kaddu et al. occurred on the trunk, as in our case. Distinctive histological features are pleomorphic, plump, spindle, and/or polyhedral cells with large hyperchromatic irregular nuclei, bizarre multinucleated cells (monster cells), and xanthomatous cells with large prominent nuclei set in a background of classic fibrous histiocytoma, including epidermal hyperplasia, grenz zone, spindle cell areas showing a storiform pattern and entrapped thickened, and hyaline collagen bundles, especially at the periphery.[11]

In our case, the histopathology report suggested cellular tumour in the subcutaneous plane with spindle cells and histiocytes arranged in a storiform pattern, fascicles and sheets. The cells showed pleomorphism and hyperchromatic nuclei. Mitotic figures were seen in 2-3/10 HPF.

CONCLUSION

In conclusion, atypical fibrous histiocytoma is an under diagnosed or a neglected entity among the cutaneous histiocytomas. Clinical suspicion, relevant investigations and proper histopathological examination is necessary to diagnose this rare variant. The prompt diagnosis and treatment help to prevent unnecessary and aggressive treatment as this is rarely malignant and rarely metastasize. The best possible treatment for this variant would be a wide local excision with clear margins.

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