A Rare Bednar Tumour Of Neck
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Citation

Abstract
Bednar tumour ( pigmented Dermatofibrosarcoma protuberans ) is a variant of dermatofibrosarcoma protuberans(DFSP)- constitute 5% of all DFSP-a relatively uncommon soft tissue neoplasm with intermediate to low grade malignancy rarely metastasizing to regional lymph nodes or distant sites but prone for local recurrence.

We report here in a case of 68year old female who presented with a huge recurrent Bednar tumour(40cm X 36cm x 26 cm) arising from neck region after a gap of five years of primary excision.

Review of previous studies and histopathological examination of recurrent lesion was suggestive of Bednar tumour. We performed a wide excision with 3 cm free skin margins. Soft tissue reconstruction was done to close the defect at the resected area.Recognition of this tumour is important because of excellent prognosis after adequate surgical excision.

INTRODUCTION
Pigmented dermatofibrosarcoma protuberans or previously known as Pigmented storiform neurofibroma is a rare variant of DFSP ,described by BEDNAR in 19571.The histological picture shows melanin laden dendritic cells with spindle shaped cells in storiform pattern.The tumour mostly occurs on trunk,upper and lower extremities,few in head and neck region2..It is a slow growing,locally invasive-painless cutaneous multilobulated lesion, burn or surgical scar3 at the initial stage often the cause.

It has been shown that Giant cell Fibroblastoma has a close histogenic relationship to Dermatofibrosarcoma protuberans4 and it has been reported that Giant cell fibroblastoma can transform into DFSP5-7.We present here a case of massive Bednar tumour of neck region presenting five years after initial excision.

CASE REPORT
A 68 year old female was admitted with a huge painless nodular tumour over the neck region. She gives history of insect bite at the same spot eight years back following which a tumour developed over next one year slowly growing upto four cm in diameter. The lesion was excised under local anaesthesia and histological diagnosis revealed Dermatofibroma. She had another recurrence of the tumour two year following the first excision which she neglected and the tumour gradually progressed to a very large size.

The patient denied fever , chills, bony pain or any other constitutional symptoms. On physical examination,patient was not cachectic with no palpable lymphadenopathy.Respiratory, CVS and GI exam was within normal limits.

Skin examination revealed 40 x 34 cm exophytic mass over neck region with multiple soft protuberances of varying size none exceeding 8 cm,soft in consistency with thin skin cover over certain areas with ulceration. The nodules appeared fixed to the overlying skin but were mobile over the deep tissue. There was no other positive finding on complete head and neck examination and there was no other swelling elsewhere in the body.
Lab. Findings were within the normal limits. Chest X-ray was WNL.

Computed tomography (CT) revealed no invasion of the mass into deeper structures.

A wide and deep excision with 3 cm margins of normal tissue all around the lesion was performed. Grossly, the resected specimen measured 40 X 36 X 26 cm diameter with multiple discrete nodular mass, the greatest being 8 cm in diameter. The nodules were well circumscribed, had a firm consistency and showed a whorled pattern on cut section.

On histologic examination, they were found to be composed of a uniform population of fibroblast like cells, arranged in a storiform pattern. The cells had hyperchromatic oval to spindle shaped nuclei with low mitotic activity. The striking feature was presence of bipolar and multipolar dendritic cells with tentacle like processes emanating from nucleus.

Immunohistochemical study showed positive reaction to CD 34 and VIMENTIN and negative for protein S-100 but melanin containing cells showed positive for protein S-100.

The resected specimen showed tumour free margins. Soft tissue reconstruction was done at the resected site of tumour.
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Figure 3
Neoplasia occupying the dermis and compose of fusiform cells and cells containing melanin

Figure 4
Melanin pigment seen in cells of dermis and Subcutaneous layer

Figure 5
Immunohistochemistry—Positivity for antibody against CD 34, which confirmed the diagnosis

Retrograde histological re-examination of the specimen obtained from the second excision of the recurrent tumour showed high expression of CD34 and revealed present tumour was actually a recurrence of this tumour.

The wound healed without any complication and the post operative course was uneventful, with no paraesthesia or skin sloughing at the reconstruction site.

DISCUSSION

The case presented here was DFSP, which was initially mistaken to be dermatofibroma. Dermatofibromas composed predominantly of fibroblasts extending into the subcutaneous tissue are difficult to distinguish from dermatofibrosarcoma protuberans though the patterns are different.

Bednar tumour account for less than 5% of all DFSP and more common in Black male population. Different theories have been given regarding origin of pigment laden cells. Dual cell origin suggests CD 34
positive spindle cells of Mesenchymal and pigmented cells of neuroectodermal origin10.

In the case reported here correct clinical diagnosis was not reached initially but only later on recurrence after subjecting to histopathological and immunohistochemical studies. Condensation of connective tissue at the periphery may give a false appearance of encapsulation but tumour may extend well beyond margins11. We need to be aware of this condition and confirm histological diagnosis before excision.

Recurrence are due to inadequate excision with tumour extending to deep resection margin. With recurrence the lesion becomes less well differentiated13 and chances of metastasis increases. Though rare, dissemination occurs by hematogenous route and rarely lymphatically. The principal site being lungs though bones, liver, pancreas, stomach, intestine, thyroid and brain may be involved.

Surgery with a safety margin of 3 cm including the underlying fascia serves good with Computerized tomogram helpful in deciding the line of incision to prevent recurrence or metastasis. Mohs micrographic surgery14 has maximum oncologic effectiveness and is the accepted treatment of choice.

Chemotherapy is not used in treatment while radiotherapy can be used as adjunct to surgery in cases with positive resection margins. Molecular targeted therapy and Imatinib may provide alternative treatment to unresectable tumours or adjunctive treatment in addition to surgery.

In short, Bednar tumour is a rare tumour. Painless, cutaneous and multilobulated lesion should arouse suspicion of this tumour and core or incision biopsy should aim at pre-operative histologic diagnosis. Adequate excision will avoid recurrence and excellent prognosis.

References
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