IJNRD.ORG ISSN : 2456-4184
INTERNATIONAL JOURNAL OF NOVEL RESEARCH AND DEVELOPMENT (IJNRD) | IJNRD.ORG
An International Open Access, Peer-reviewed, Refereed Journal

Pigmented Dermatofibrosarcoma Protruberans: A Rare Case Report

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INTRODUCTION :

Dermatofibrosarcoma protuberans (DFSP) is a cutaneous fibrous neoplasm of intermedi- ate malignant potential. It is composed of a monomorphic population of spindle shaped cells in a characteristic storiform pattern. It affects individuals in 3rd and 4th decades of life predominantly, although cases have been reported in infants also. Cases have been described in all ethnic groups with a slight predominance among blacks.
Bednar tumor, which was formerly known as storiform pigmented neurofibroma, was later established to be a pigmented variant of DFSP because of the histological and cytogenetic similarities between the two. Bednar tumor differs from a classical case of DFSP due to the presence of pigment laden dendritic cells amidst the fusiform cells that are characteristic of DFSP. Bednar tumor can rarely show malignant transformation and can also rarely recur.

CASE REPORT :

A 38 year old female presented with complaints of a mass over lateral aspect of left lower leg just above the lateral malleolus. The mass was not causing pain or discomfort to the patient. On examination the mass was found to be a pigmented nodule measuring 2 x 1.5cm(Fig A). Clinical diagnosis of Fibroma-left leg was made. There were no similar lesions elsewhere in the body. Grossly, the mass was a skin covered nodule measuring 2 x 1 x 0.5 cm. Cut surface of the mass showed brownish to yellowish areas(Fig B).

Microscopically, sections studied showed skin with subcutaneous tissue .(Fig C) Epidermis showed mild acanthosis and mild increase in melanin pigmentation(Fig D). Dermis shows an uncircumscribed highly cellular tumor mass, comprised of a single population of mono- nuclear spindle shaped cells arranged in storiform pattern and exhibiting mild atypia (Fig E). Amidst this, numerous heavily pigmented cells were seen (Fig F). On high power, there were brownish black melanin pigment laden round to polygonal cells among the spindle neoplastic elements (Fig G). Spindle cells show moderate amount of eosinophilic cytoplasm with hyperchromatic nuclei. Few mitotic figures were also noted along with lymphocytic infiltrate. Some multinucleated giant cells were also seen, with a few showing pig- mentation within the giant cells (Fig H). No haemorrhage or necrosis is seen. Immunohisto- chemical study showed positivity of pigmented cells with HMB 45 (Fig 1) and S-100 (Fig. J) and tumor cells show positivity for CD 34(Fig K).

Fig A: Gross appearance of specimen

Fig B:Cut section of mass

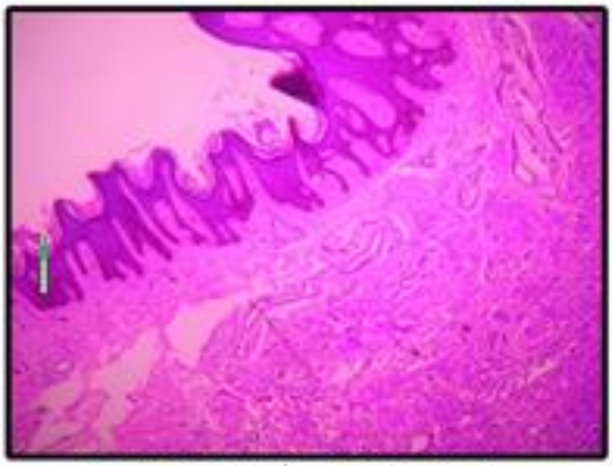

Fig C:Epidermis with underlying dermis showing tumor mass(H & E, 100X)

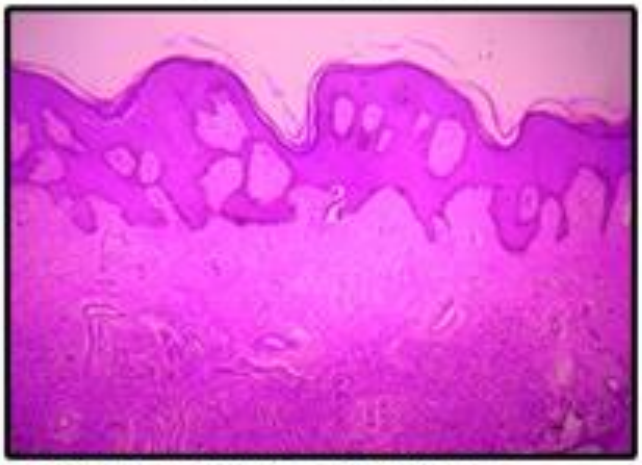

Fig D: Epidermis showing mild acanthosis and mild increase in melanin pigmentation. (H & E, 100X)

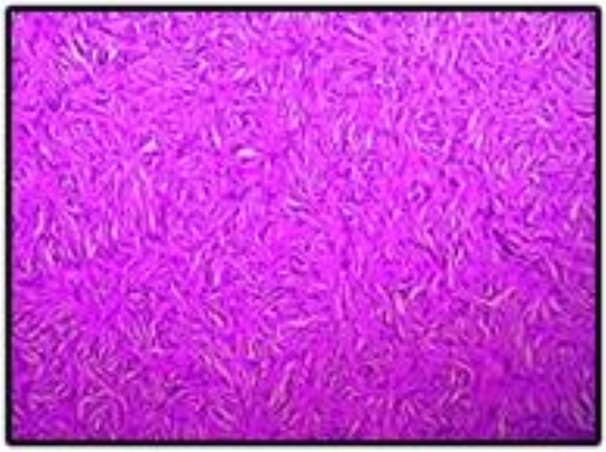

Fig E: Tumor mass composed of spindle shaped cells in storiform pattern (H & E, 400X)

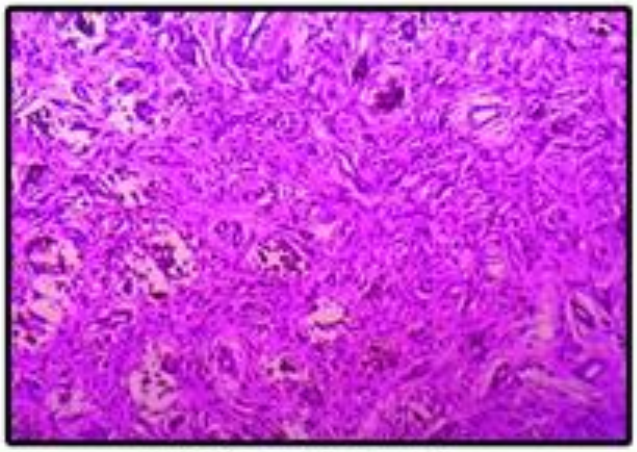

Fig F: Pigmented cells within tumor cells (H & E, 400X)

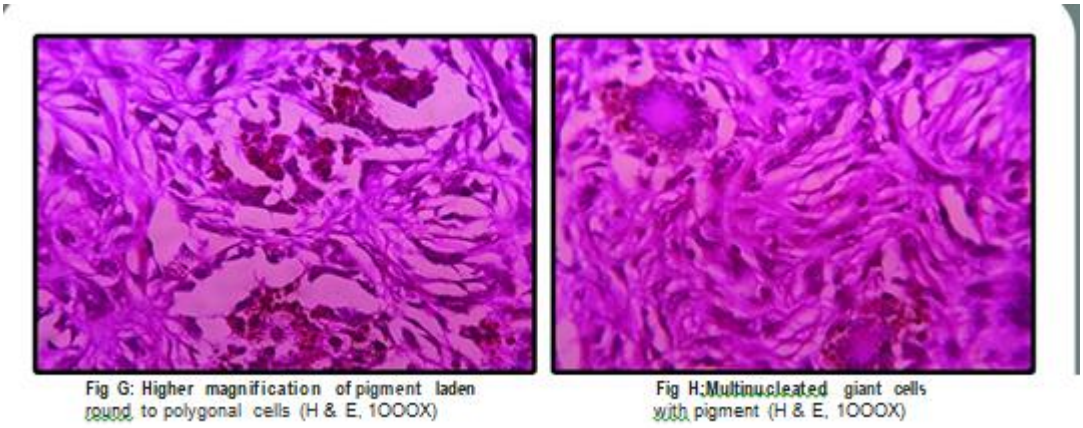

Fig G: Higher magnification of pigment laden round to polygonal cells (H & E, 1OOOX)

Fig H:Multinucleated giant cells with pigment (H & E, 1OOOX)

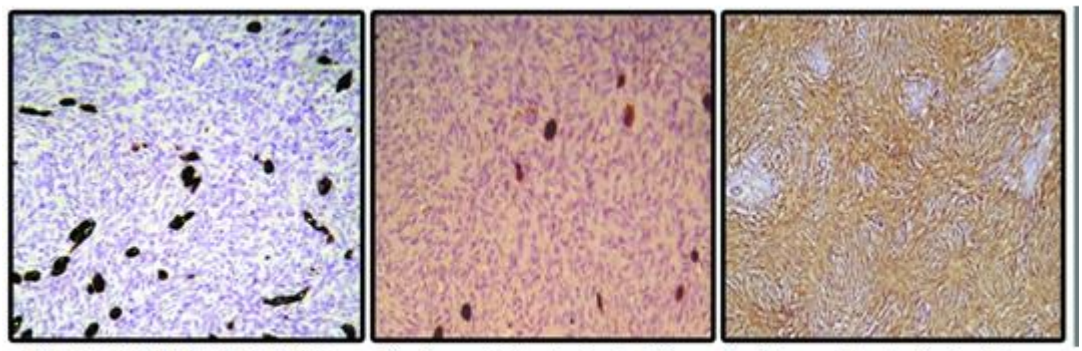

Fig I:Immunohistochemical staining shows pigment cells with positivity for HMB45(400X)

Fig J:Immunohistochemical staining shows pigment cells with positivity for S100(400X)

Fig K:Immunohistochemical staining shows tumor cells staining positive for CD34(400X)

DISCUSSION

DFSPs are uncommon fibrous neoplasms constituting around 0.1% of skin tumors. Bednar tumor is a rare pigmented variant of DFSP which was first described in 1957 and t constitutes 1-5% of all DFSPs. Several theories have been proposed for the histogenesis of Bednar tumor. One theory suggests that the tumors are of neuroectodermal origin due to the presence of dendritic melanocytes. Cells suggestive of Schwannian differentiation also support this theory. Immunohistochemical studies show the cell of origin to be a neuromesenchymal cell. Another theory attributes the origin of this tumor to various kinds of local trauma including previous burns, vaccination scars or insect bites. Our case sup- ports the former theory.

The tumor most frequently presents as pigmented nodules with irregular surface over the skin or even deep rooted within the subcutaneous tissue. Most common site of presentation is the trunk region, followed by upper and lower extremities, and then by head and neck region.

Main differential diagnoses of Bednar tumor include other pigmented spindle cell lesions like desmoplastic malignant melanoma, neurocristic cutaneous hamartoma, pigmented neurofibroma, and psammomatous melanotic schwannoma. Pigmented neurofibroma can be differentiated from Bednar tumor by the extensive storiform growth and strong positivity for CD34 in Bednar tumor. Psammomatous melanotic schwannoma will present as a circumscribed, heavily pigmented mass with psammoma bodies and tumor cell show positivity for S100 and negativity for CD34. Desmoplastic malignant melanoma can be differentiated from Bednar tumor by the presence of junctional activity and neurotropism.

Here we reported case of a 38 year old female who presented with a mass over her left leg. As the classical features of DFSP were seen in histopathology, the differentials were ruled out. Bednar tumor differs from the classical case of DFSP due to the presence of melanin containing dendritic cells and it gives focal positivity for HMB45, Melan A and tyrosinase. Pigmented cells also give positivity for S-100. Tumour cells give positivity for CD34 and vimentin.

CONCLUSION

DFSP is a rare fibrous neoplasm of intermediate malignant potential, composed of monomorphic spindle shaped cells in storiform pattern. Bednar tumor is an uncommon pigmented variant of DFSP. It can rarely undergo malignant transformation in the form of fibrosarcoma with recurrence and metastasis. Therefore frequent review and follow-up is recommended as we are doing for this patient.

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