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PIGMENTED DERMATOFIBROSARCOMA PROTRUBERANCE (BEDNAR'S TUMOR)-A RARE ENTITY.

Medical Science

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ABSTRACT

Bednar's tumor is a rare skin neoplasm, considered to be a pigmented variant of dermatofibrosarcoma protuberans. It accounts for approximately 1-5% of all cases of dermatofibrosarcoma protuberans (DFSP). This tumor is of unexplained histogenesis but it may be related to remnants of embryonic mammary tissue or to various kinds of local trauma such as burns, vaccination scars and insect bites. Majority of lesions are located on the trunk, and the remainder are more or less equally distributed in the upper and the lower extremities and the head and neck and present as exophytic, multinodular neoplasms of the dermis or subcutaneous tissue. Microscopically the lesion is characterized by spindled cells arranged in a repetitive storiform pattern admixed with a small population of melanin-containing dendritic cells.. Bednar's tumor is locally aggressive and recurrences are frequent, but metastases are rare. The most appropriate therapeutic procedure is Mohs' micrographic surgery. The case report presents a 35-year-old patient with this rare neoplasm in which the diagnosis was established through histopathologic examination and immunohistochemical study.

KEYWORDS

Bednar's tumour, DFSP, dendritic cells,melanin.

INTRODUCTION:

Bednar's tumors are rare; they were described by Bednar in 1957.[1]. Their incidence is approximately 1-5% of all cases of DFSP.[2] .They are considered to be a pigmented variant of dermatofibrosarcoma protuberans (DFSP), and differ from conventional DFSP by the presence, of fusiform dendritic cells containing melanin, dispersed amidst the spindle cells characteristic of DFSP on histopathology. They are usually seen in the third and fourth decades of life, however they may also occur in infancy [3]. They have been described in all ethnic groups, but with a predominance in Blacks[4]. The common site of origin is the trunk, especially the back and shoulders. Most tumors present as slow-growing, wide-based, protruding masses or plaques in the dermis and subcutaneous tissue. Clinically the tumor in our case has been described to be recurrent (with a previous history of operation) multinodular swelling, having irregular surface and firm nodules growing deep within the subcutaneous tissue. Diagnosis was established by histopathologic exam and immunohistochemical study.

CASE REPORT:

A 35-year-old man had a large multinodular swelling on the left lateral flank for the last 5 years. The swelling was excised in the past (6 years back) on the same site . The histopathological records of which were not available. The patient now presented to us with swelling which was 8×3×3 cms, grayish black in color, multinodular, nontender, not fixed to the underlying tissue and slightly mobile [Figure 1]. There were no satellite lesions. A fine-needle aspiration of the lesion was performed and Giemsa staining done. The cytologic features revealed a spindle cell lesion obscured by pigment. The tumor was resected along with fascia and a clinically tumor-free margin. The sample was sent for histopathological study to our department. Gross examination showed a multinodular swelling grayish brown in color measuring 8x3x3.On c/s it was solid and homogenous. The histopathologic examination revealed proliferating spindle cells arranged in repetitive storiform pattern. On higher magnification, the spindle cells were uniform appearing with a moderate amount of eosinophillic cytoplasm and plump nuclei with tapering edges.. These pleomorphic cells were seen infiltrating into the underlying dermis around appendages and into the subcutaneous tissue.. The cells showed coarse brown black pigment obscuring the nuclei. No areas of necrosis or hemorrhage were noted Fig 2(a),(b)(c). The above-mentioned features lead to the diagnosis of Bednar's tumor. The immunohistochemistry showed positivity for CD34. A final diagnosis of Bednar tumor was made.

DISCUSSION:

Initially designated as "storiform neurofibroma" by Bednar in 1957, this variant of dermatofibrosarcoma protuberans contains abundant melanotic pigment [1].The biological behavior is that of intermediate malignancy. The lesions present a slow growth, over a period of months or years. The published cases generally report: the lesion clinically as plaque lesions of an erythematous-bluish or brownish color, with a smooth or irregular surface. In some cases the lesions are exophytic, nodular, multilobular and of firm consistence as in our case. Their invasive growth into the dermis, may reach the subcutaneous strata, fascia and profound musculature, in a manner similar to that of dermatofibrosarcoma protuberans.

The histogenesis of this peculiar tumour is still under debate, it may be related to remnants of embryonic mammary tissue or to various kinds of local traumas, such as previous burns, vaccination scars, insect bites or vaccination (BCG) [5]. Neuro-ectodermal differentiation or melanocytic colonization are the two proposed theories for histogenesis for the Bednar tumor. It has also been reported in association of dermal melanocytosis (nevus of Ito), and based on the immunohistochemistry, the cell of origin is thought to be a neuromesenchymal cell. Three populations of cells have been identified in Bednar tumors in studies by electron microscopy. Most of the cells are represented by fibroblasts. The second cellular population exhibits elongated, fine elongations, partially or completely enclosed in basal membrane. The third population consists of dendritic cells containing melanosomes and premelanosome.

In immunohistochemical studies most of the tumor cells exhibit a positive reaction to CD 34 and vimentin, and are negative to neuron-specific enolase, HMB-45 and protein S-100. However, those cells containing melanin may react positively to protein S-100 and vimentin.[6].CD34 was diffusely positive in our case.

The pigment-laden dendritic cells has to be distinguished from other pigmented cutaneous spindle cell lesions like pigmented neurofibroma, psammomatous melanotic schwannoma, neurocristic cutaneous hamartoma and desmoplastic malignant melanoma. However, our case had classic features of DFSP, and the above lesions were ruled out by histopathology itself.

The treatment of choice is Mohs' micrographic surgery, because the principal source of recurrence of the tumor seems to be from cellular

groups located deep within the subcutaneous cellular tissue, fascia and muscular bundles, that might not be excised by conventional surgical techniques.

Bednar tumor can rarely undergo malignant transformation in form of fibrosarcoma with repeated recurrences and distant metastasis. Metastasis is rare and late and dissemination may occur via the hematogenous route and rarely lymphatic. The principle sites of metastasis are lungs, bones, liver, pancreas, stomach, intestine, thyroid and brain.[7] Hence, a close follow-up of these cases is always necessary.

CONCLUSION:

Bednar tumor poses a diagnostic challenge to the dermatologist in terms of its recognition. The clinical aspect is most commonly suggestive of a diagnosis of melanoma or residual inflammatory lesion. So histopathologic examination complemented by immunohistochemical study is fundamental for diagnosis. To conclude, though rare, it is important for the histopathologists to be aware and recognize this unusual entity and distinguish it from other pigmented spindle cell lesions.

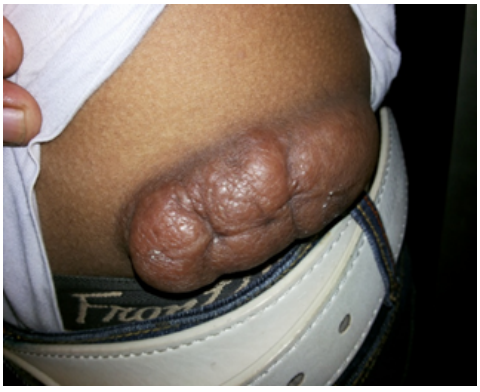

Fig 1- Multilobular exophytic growth measuring 8x3x3 on lateral left side flank.

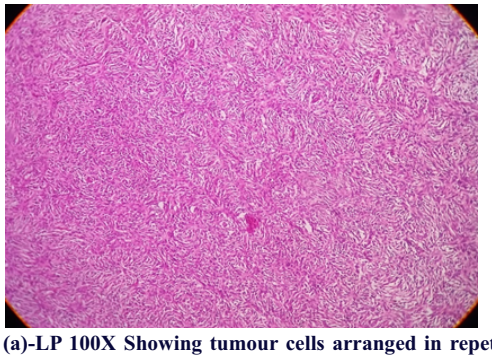

Fig 2 (a)-LP 100X Showing tumour cells arranged in repetitive storiform pattern .

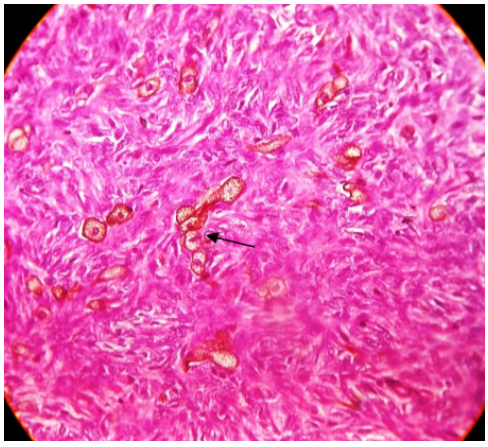

Fig 2 (b)-HP 400X- Showing fusiform melanin containing dendritic cells admist spindle cells.(arrow)

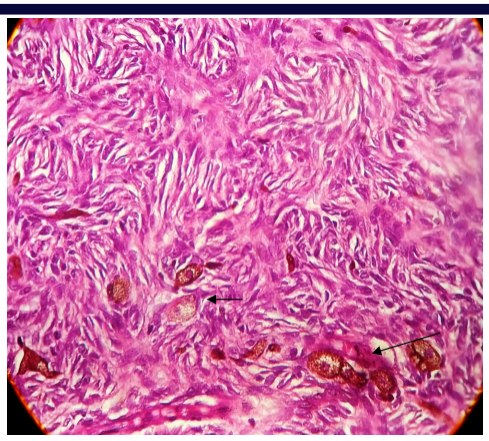

Fig 2 (c)-HP400X- Showing fusiform melanin containing dendritic cells admist spindle cells.(arrow)

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