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A Rare Soft Tissue Tumour.

Namasivayam T*, Karthikeyan CT, Francis RNM, Darwin P, and Inbaselvi P.

Sree Balaji Medical College & Hospital, Bharath University, Chrompet, Chennai – 600044, Tamil Nadu, India.

ABSTRACT

Bednar tumor is a rare skin neoplasm, considered to be a pigmented variant of derma to fibro sarcoma pro tuber ans[1]. Diagnosis is confirmed through his to pathologic examination and immune his to chemical evaluation. Bednar tumor is aggressive locally and recurrences are frequent, but metastases are rare. The most appropriate therapeutic procedure is Mohs' micrographic surgery[2]. The case report presents a 66-year-old patient with this rare neoplasm in which the diagnosis was established through his to pathologic examination and immune his to chemical study.

Keywords: soft tissue, tumor, skin neoplasm

**Corresponding author*

INTRODUCTION

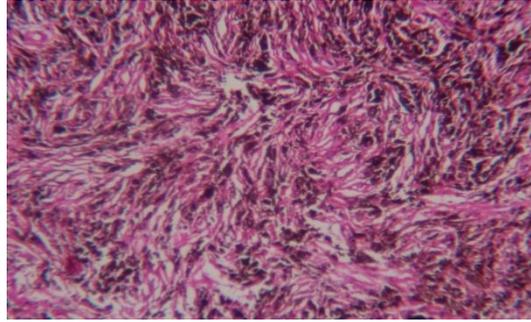
Bednar tumors are rare; they were described by Bednar in 1957[1]. They are considered to be a pigmented variant of derma to fibro sarcoma pro tuberans (DFSP), and differ from DFSP by the presence, in the his to pathologic exam, of dendritic cells containing melanin, dispersed amidst the fusi form cells characteristic of DFSP. They involve individuals predominantly in the third and fourth decades of life however they may also occur in infancy.[3,4]

CASE REPORT

A 66 year-old female presented with complaint of swelling in the right log in for 7 months duration . The swelling is gradually increasing in size to reach the present size. There was no H/O trauma, fever, pain. O/E a single swelling of size 8×8×7 cm in Right loin, surface is smooth, firm in consistency, edges of the swelling are well defined. Skin over the swelling is stretched. No local warmth / tenderness/ pulsations present over the swelling.

On further investigating the patient, MRI showed a well-defined bi-loculated cystic lesion with thick irregular walls in subcutaneous plane, with possibilities of soft tissue neoplasm, epidermal inclusion cyst or hematoma. True-cut biopsy was done, but aspirated only blood stained fluid, which was cytologically analyzed and negative for malignant cells. So wide local excision biopsy was done A blackish soft tissue mass, with cross section showing hemorrhagic areas, was sent for His to pathologic examination. The report came as BEDNAR TUMOR (Derma to fibro sarcoma pro tuber ans).





Spindle cell lesion composed of elongated cells with pleomorphic nuclei arranged in fascicles and interlacing bundles. Many tumour cells show brownish black pigmentation

DISCUSSION

Initially designated "*stori form* neuro fibroma" by Bednar in 1957, this variant of derma to fibro sarcoma protuberans contains abundant melanotic pigment. The Bednar tumor is rare.

It has been described in all ethnic groups however it is generally more prevalent in blacks. It usually occurs in the third and fourth decades of life, but cases have been described in children and neonates, including the occurrence of a congenital Bednar tumor.

The lesions present a slow growth, over a period of months or years. The most frequent location is in the trunk. Other areas that may be involved are: upper and lower members, or the head and neck.

The clinical aspect is most commonly suggestive of a diagnosis of melanoma or residual inflammatory lesion, therefore his to pathologic and immune his to chemical exams are necessary for the correct diagnosis. In immune his to chemical 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. [5]

A Bednar tumor is aggressive and locally invasive, with a pronounced tendency to local recurrence. Metastases are rare and late, and dissemination may occur preferentially via hematogenic means but rarely lymphatically.[6] The principal site of metastasis is the lung, however the bones, liver, pancreas, stomach, intestine, thyroid and brain may be involved the treatment of choice is Mohs' micrographic surgery.[7]

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