

Fine Needle Aspiration Cytology of Pigmented Dermatofibrosarcoma Protuberance (Bednar Tumor) & it's Pathogenesis

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Abstract: Dermatofibrosarcoma protuberance is a soft tissue tumor of low malignant potential. At times it is associated with pigment deposits which are then it is termed as Bednar tumor. Only a few Fine Needle Aspiration Cytology studies of Bednar Tumor have been reported. We share our findings of Fine Needle Aspiration Cytology in a 30 year old man with Bednar tumor of scalp and multiple enlarged cervical lymph nodes. Our smears revealed scattered atypical large round to oval cells with scant cytoplasm. Some cells' cytoplasm contained abundant melanin pigment which also resulted in dirty black pigmented background. The tissue sections showed melanin in both spindle shaped cells and rounded cells. At places it seemed that heavily pigmented cells became oval and globular and necrotic. The melanin pigment was extended into the cellular processes. The overlying skin epidermis shows focal kilocytotic type change. The tissue sections showed positivity for CD 34 and protein S-100. We propose that round globular cells are derived from spindly cells after melanin synthesis and when the cytoplasm is overloaded with melanin the cells undergo necrosis. Focal kilocytotic type change in overlying epidermis deserves further investigation.

Keywords: Soft tissue tumors, Dermatofibroma, Dermatofibrosarcoma protuberance, Pigmented Dermatofibrosarcoma Protuberance, Bednar tumor, Immunohistochemistry CD34 & Protein S-100, HPV, Kilocytotic atypia, melanin

Introduction

Dermatofibrosarcoma protuberant (DFSP) is an uncommon, slow growing, intermediate grade subcutaneous tumor that rarely metastasizes. It may occur anywhere on the body but trunk and proximal extremities are common sites. Rarely does it arise in head and neck region. The patients are usually middle aged but can be infants and young children¹⁻⁵. It may invade underlying fat, fascia, muscle and bone. About 1% of these tumors may contain black-brown pigment. It's then labeled as Bednar tumor. Bednar tumor is more common among blacks.

Case Presentations

A 30 year male patient presented with a 5 cm diameter tumor on scalp for the past 7 months. He later developed multiple enlarged lymph nodes which were tender and painful.

He also noted a small swelling on his back. We performed FNAC on his scalp and left cervical lymph nodes. Diagnosis of malignant lesion was made with recommendation of biopsy. It turned out that he already had the biopsy which was diagnosed as 'Dermatofibrosarcoma Protuberance (Bednar's Tumor). We re-examined the histopathology slides as well. For immunohistochemistry the slides were first treated with hydrogen peroxide solution with phosphate buffer overnight to remove melanin from the tissue sections. Then standard immunohistochemistry procedure was followed.

Microscopic Description

The FNAC smears were riddled with dark black granules in the background which had apparently escaped through ruptured membranes of pigmented cells (Figure 1). There were many scattered atypical cells with scant cytoplasm, high N/C ratio (Figure 2). Prominent breaks and pits were noted in the nuclear membranes (Figure 3). Some cells showed molding and the nuclear chromatin was blotchy and coarsely granular (Figure 4). In some cells these granules almost completely masked the nuclei and these

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melanin granules extended into the long cellular processes (Figure 5). The tissue sections showed hyperplastic epidermis with irregular growth of rete ridges (Figure 6). The epidermis showed focal cytotoxic type nuclear changes with rectangular crumbled nuclei and perinuclear halos (Figure 7). Deep down, the reticular dermis contained a well-defined tumor composed of markedly proliferated spindle fibroblasts displaying distinct whorled pattern. Some of the spindle cells contained melanin pigment and at places these were transformed into round to oval shapes. Both spindle and round cells contained melanin pigment (Figures 8 & 9). The melanin pigment was seen in the long cellular processes of spindle cells (Figure 10). The heavily pigmented spindle cells appeared to transform into round globular pigmented cells which gradually underwent shrinkage or pyknosis (Figures 11-14). The tumor at places showed frank necrosis without presence of pigmented tumor cells. (Figure 15). Focal lymphocytic infiltrate was seen. (Figure 16). The tumor cells were positive for CD 34 and protein S-100 (Figure 17). The Prussian blue stain for iron however was negative.

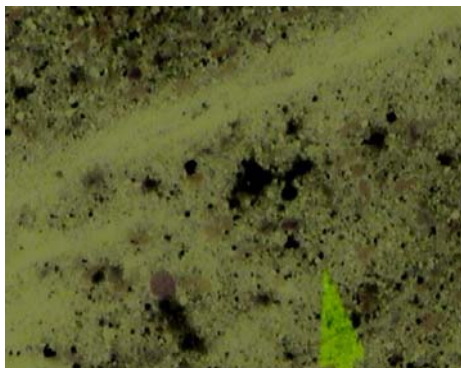


Fig.1: FNAC smears showed dirty black pigmented background (Hemacolor X 40)

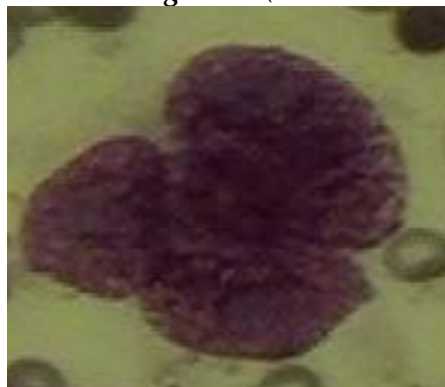


Figure 2: Nuclear membrane breaks (Hemacolor X 1000)

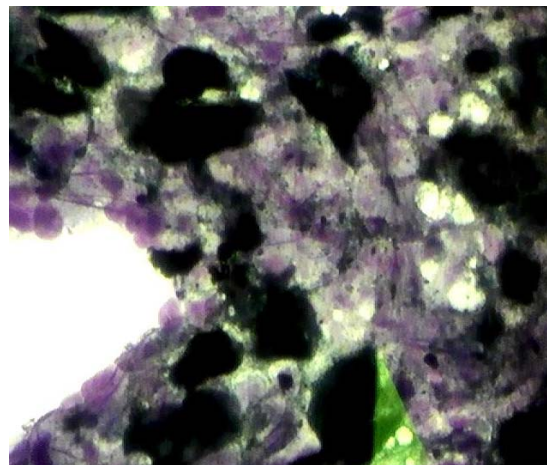


Figure 3: Heavily pigmented atypical cells (Hemacolor X 400)

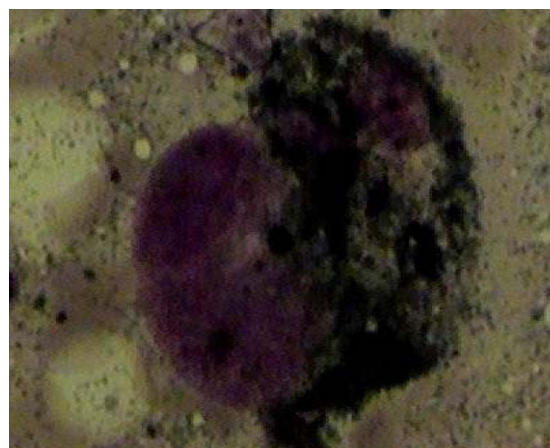


Figure 4: Two atypical cells; one molded contains abundant melanin (Hemacolor X 400)

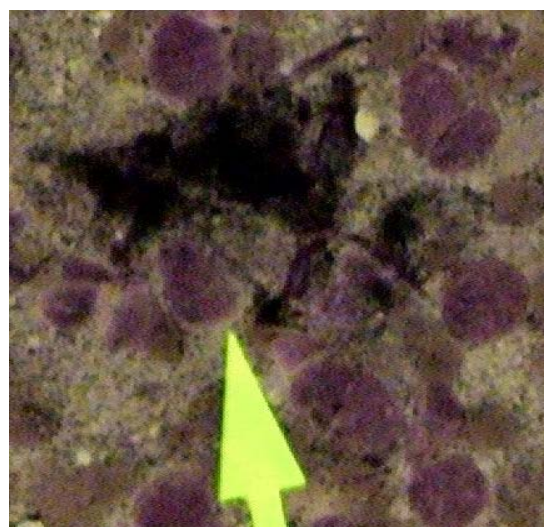


Figure 5: Pigment is seen in the cell process of the atypical cells. (Hemacolor X 400)

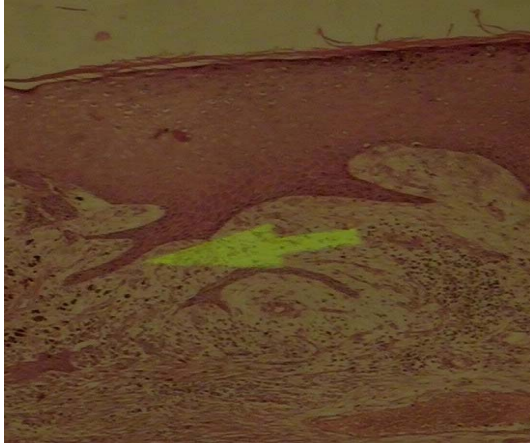


Fig.6: Hyperplastic epidermis with irregularly proliferated rete ridges. (H&E X 100)

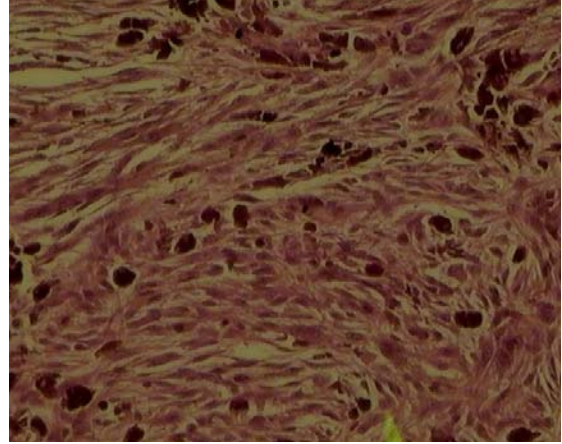


Fig 9: Amidst whorled "fibroblasts" many pigmented rounded cells (H&E X 200)

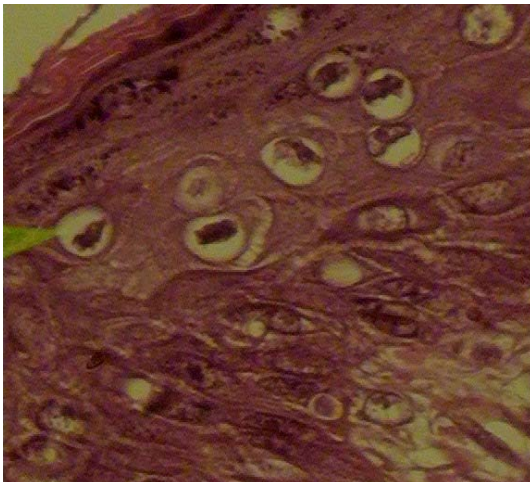


Fig 7: Epidermal cells with karyocytotic type change (arrow) (H&EX400)

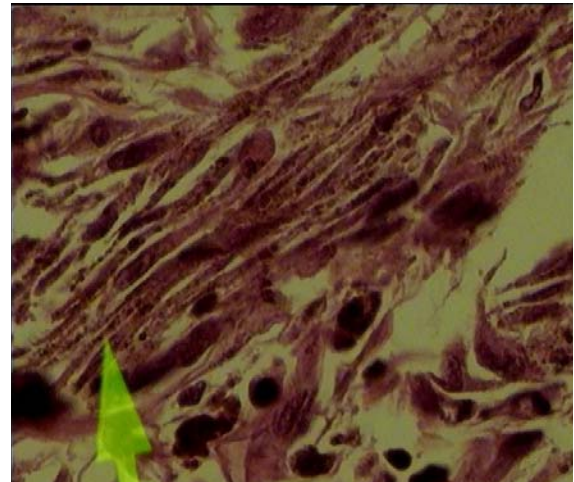


Figure 10: Melanin pigment in the long "dendritic" processes (H&E X 400)

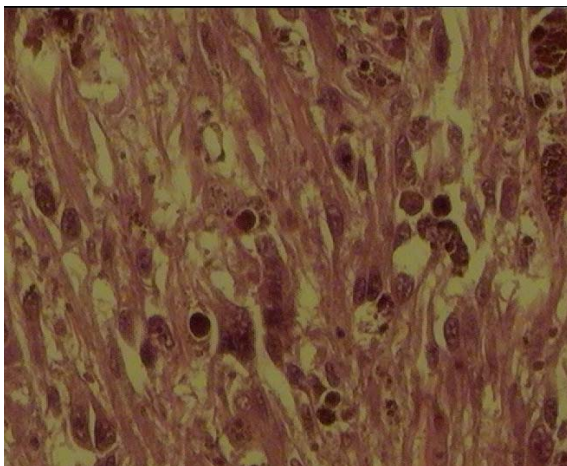


Fig 8: Pigment is present in spindle shaped cells. (H&E X 400)

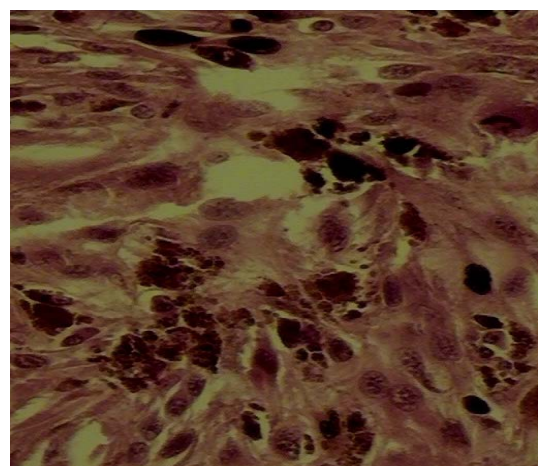


Fig 11: Heavily pigmented spindle and rounded cells (H&E X 400)

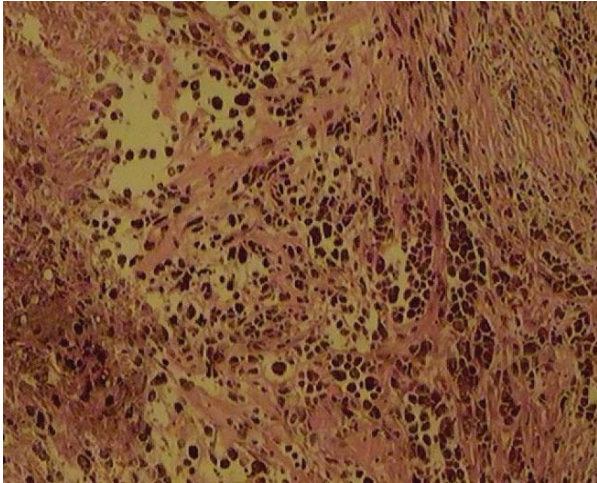


Figure 12: Pigmented rounded cells undergoing pyknosis and necrosis (H&E X 200)

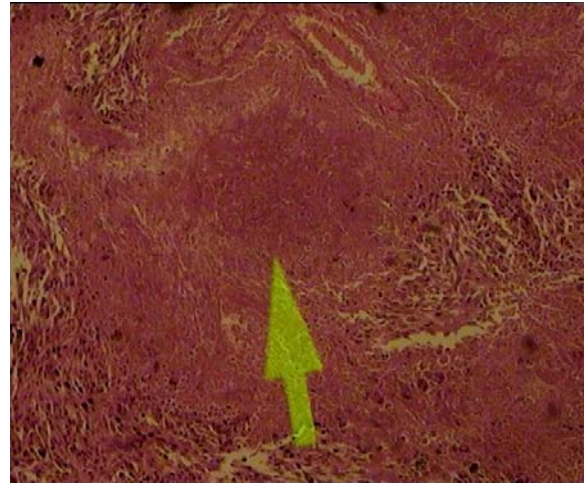


Figure 15: Areas of marked necrosis (H&E X 100)

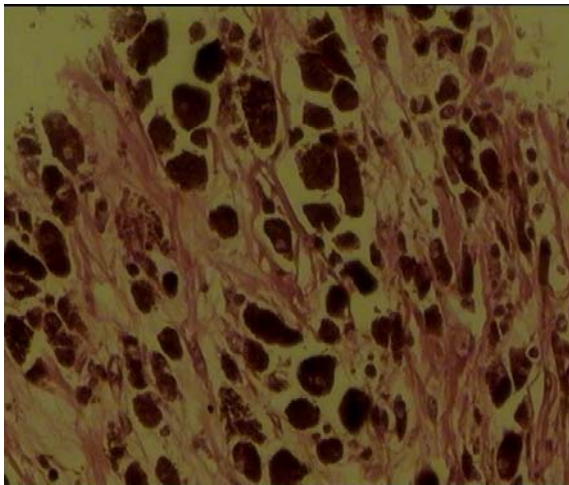


Figure 13: Shrunken heavily pigmented cells (H&E X 400)

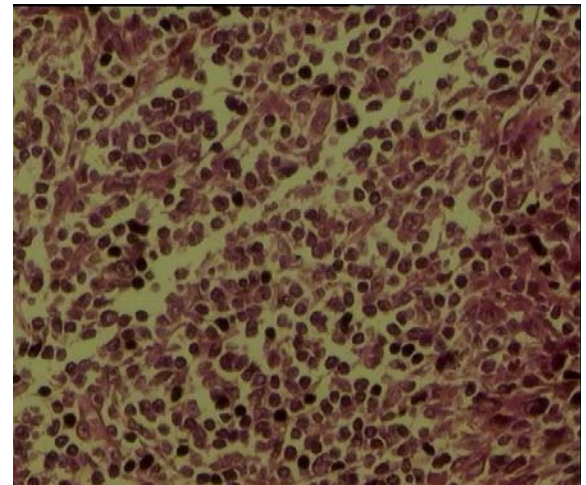


Figure 16: Focal lymphocytic infiltrate (H&E X 100)

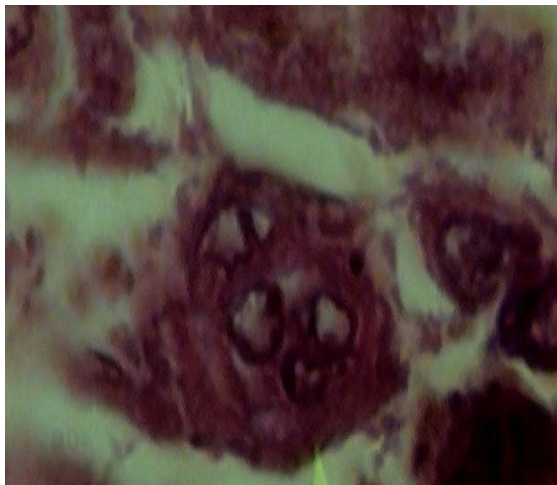


Figure 14: Necrosed crumbled cells with multinucleation. (H&E X 400)

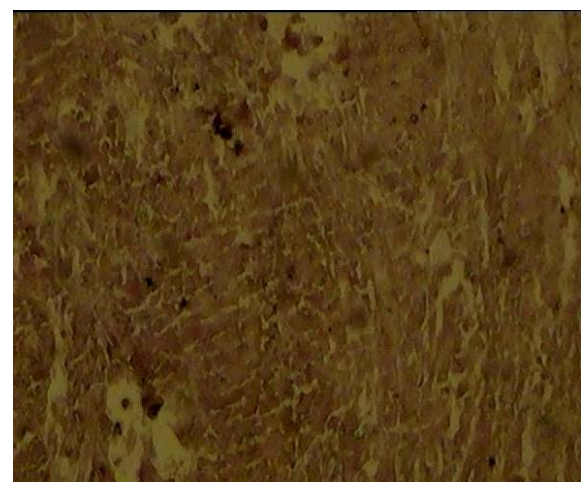


Figure 17: CD 34 diffusely positivity in the tumor cells. (Immunoperoxidase X 100)

Discussion

Fine Needle Aspiration Cytology (FNAC) was introduced in 1927 however it became popular after 4-5 decades. ⁴ It nowadays play key role in diagnosis of various epithelial, soft tissue and bone tumors and non tumor.⁵ FNAC is also used in such conditions as evaluation of spermatogenesis.⁶ Because of its numerous virtues including low cost, outpatient setting, quick results, amenable to several repeats and multiple samplings it is often used as initial diagnostic procedure. Dermatofibrosarcoma protuberant (DFSP) is a soft tissue tumor of dermis and subcutaneous tissue. It is composed of proliferated distinct spindly cells arranged in whorled pattern. Over 90% of these are CD 34 positive as opposed to dermatofibroma suggesting some fundamental difference between these tumor cells than usual fibroblasts. ⁸ Bendar tumor is pigmented variant of dermatofibrosarcoma protuberance. On FNAC of Bendar tumor one would expect to see many spindly cells along with some rounded pigmented cells! We however saw much larger number of pigmented round to oval cells with scant cytoplasm instead of spindly cells! Perhaps these round cells represent transformed spindly cells. Some of these subsequently undergo necrosis due to heavy melanin deposits in the cytoplasm. It seems these rounded or globular cells are not as cohesive as spindly cells and therefore appear in larger numbers in FNAC smear than spindly cells. This may pose problem for the pathologist as he may not consider Bendar tumor in differential diagnosis on FNAC due to paucity of characteristic spindly cells as was the case here. Heavy pigmentation would lead to the inclusion of melanoma in differential diagnosis. However prominent eosinophilic nucleoli were not seen in these cells.

Ding et all considered these tumor cells to be of fibroblastic origin. ⁴ Our case raises the possibility of mutations leading to gene rearrangement and/or activation in these fibroblasts to produce melanin. On one hand we observed long

“dendritic processes” containing melanin pigment and on the other hand pigmented fibroblastic type cells transforming into round globular cell.

We do not agree with the conception that pigmented cells are not neoplastic, we rather suggest that these cells are transformed spindly cells which later undergo necrosis due to heavy cytoplasmic melanin and release melanin pigment due o fragility as seen on FNAC smears. We conclude that Bendar tumor is a peculiar lesion with possible gene rearrangement or gene activation leading to melanin formation and transformation of spindly cells into globular cells which in turn may cause degeneration and necrosis of the pigmented cells. Presence of kilocytotic type changes in the epidermis raises the possibility of its link with Human papilloma virus. Further work is required.

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- D. Manuscript Writing
- E. Critical Review
- F. Facilitated for Reagents/Material/ Analysis