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A Case Report On Pigmented Dermatofibrosarcoma Protuberance (DFSP)

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***Abstract*: Bednar tumor is a rare skin cancer, also considered as pigmented type of dermatofibrosarcoma protuberans. Diagnosis is confirmed through histopathologic examination and immunohistochemical evaluation. Bednar tumor is aggressive locally and recurrences is very often, but metastases chances are less. The most appropriate therapy procedure is micrographic surgery. The case report presents a 24 year old patient with this rare tumor in which the diagnosis was established through histopathologic examination and immunohistochemical study. The importance is emphasized for oncologists to be keen for diagnostic suspicion and have available the necessary facts to confirm the diagnosis in order to adopt the best therapy.**

***Keywords*: dermatofibrosarcoma protuberance (DFSP), skin cancer, surgery, micrographic surgery.**

I. INTRODUCTION

Pigmented dermatofibrosarcoma protuberance (DFSP), also called as Bednar tumor, is a rare type of dermatofibrosarcoma protuberance (DFSP), described by Bednar in 1957. The lesion comprised of spindle cells arranged in a storiform pattern with elongated nuclei and a scant to moderate amount of cytoplasm. Mitotic activity was sparse. The lesion was seen move into the sub-cutaneous tissue. By doing immunological test for CD34 and factor XIIIa, the spindle cells show positive reactions for CD 34, staining for factor XIIIa has no contribution. Approximately 85% to 90% of tumors are low-grade, whereas others contain a high-grade fibrosarcomatous component. Less than 2% of cases, DFSP metastasizes and becomes lifethreatening. More than 95% of DFSP present on the 17q22 and 22q13 chromosomal regions leading to fusion of COL1A1 and PDGFB genes. Transfection studies suggest that PDGFB could act as a mitogen for tumor cells, leading to platelet-derived growth factor (PDGF) receptor activation.

II. CASE STUDY

The patient was a 24 year old male resident in india. He treated in oncology department, complaining of a "tumor on die of fore head" that appeared three years before. The lesion was symptomatic and he suggested for medical investigation after permited by his family. At dermatologic exam a lesion was observed in plaque form with very well defined margins, protuberanes; smooth surface and emerged in the lateral portion of the head, with a diameter of approximately 4.5 cm, located in the side region of head as described in Figure 1and 2 . The patient presented a good general state of health and lymphoadenomegaly or visceromegaly were not found by palpation. Biopsy of the lesion was accomplished by a 4 mm punch for histopathologic and immunohistochemical study. The histopathologic exam revealed tumor covering dermis and subcutaneous cellular tissue, composed of sindle formed cells with large nuclei, arranged in irregular bundles that are interconnected and, amidst these, spindle shaped cells containing melanotic pigment as shown in Figures 3 and 4. The panel of antibodies used in the immunohistochemical study by avidin-biotin peroxidase explain a positive reaction to CD 34. Total surgical exeresis of the lesion will performed with 6 cm wide lateral margins, corresponding to the margins in which the material obtained was histologically free from tumoral cells.

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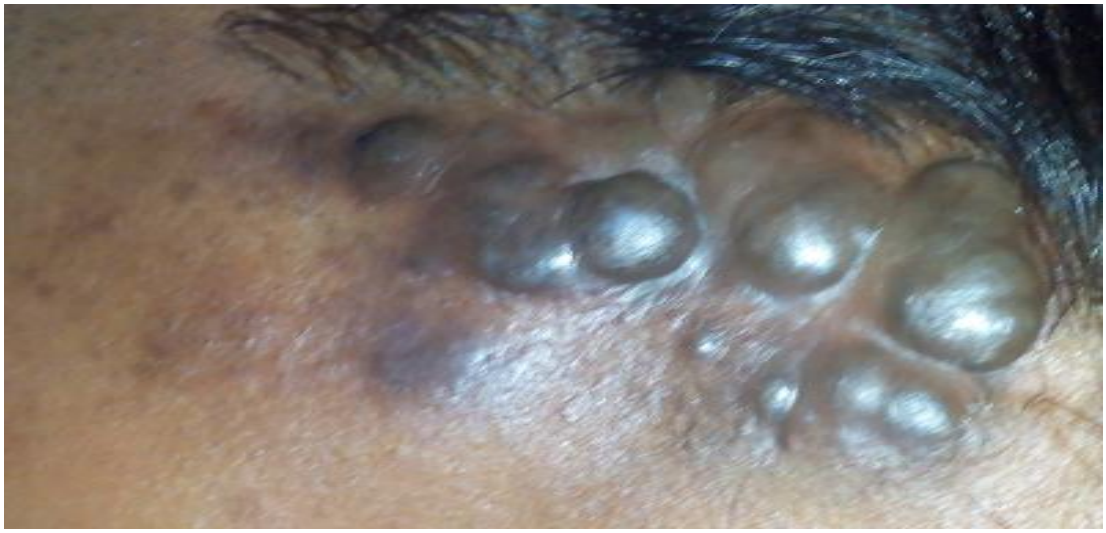
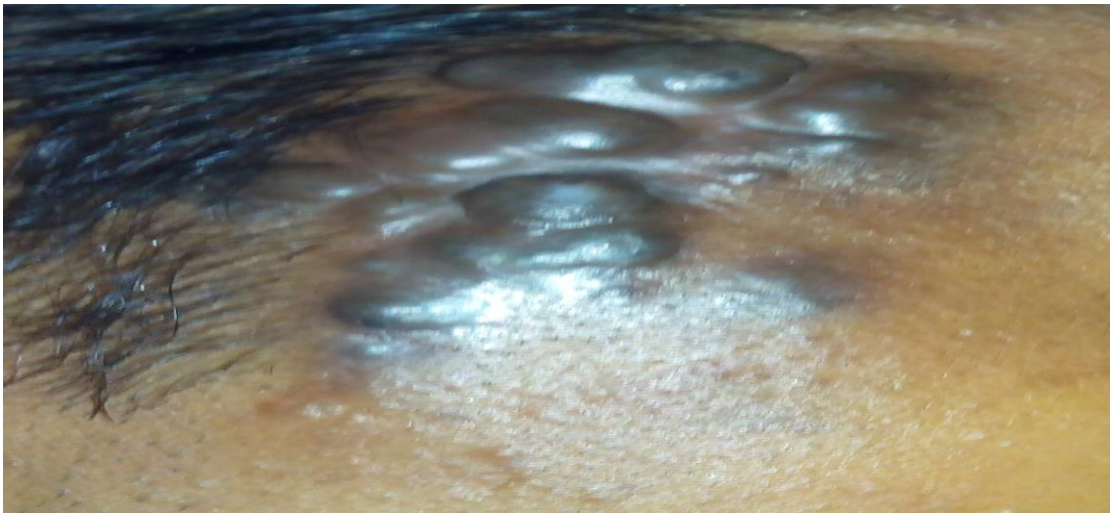
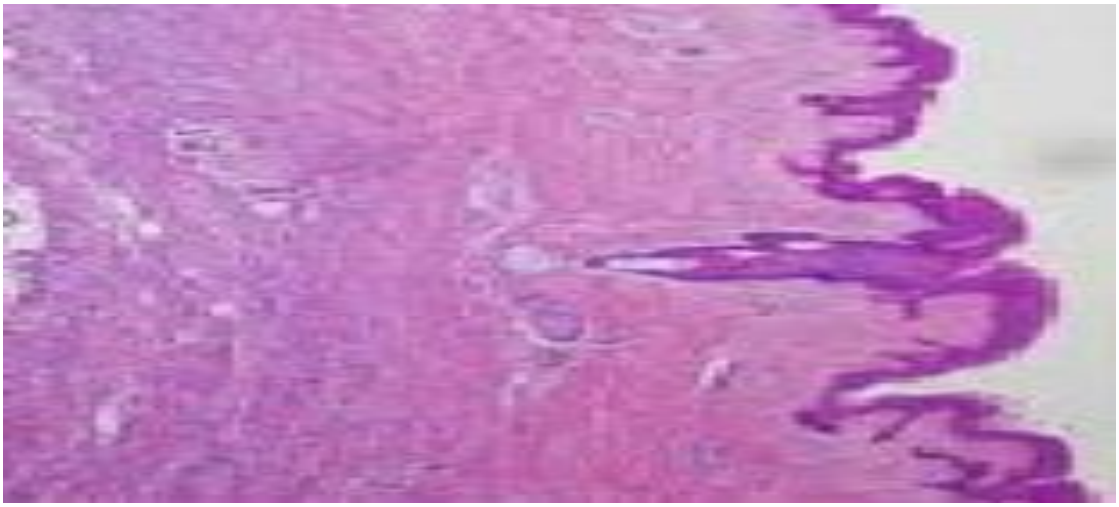


[Fig – 1: patient with bednar tumor at right lateral side of forehead, in horizental view]



[Fig – 2: patient with bednar tumor shown in actual vertical view]



[Fig-3: biopsy study of the sample taken by lesion at the area of fore head where bednar is detected]

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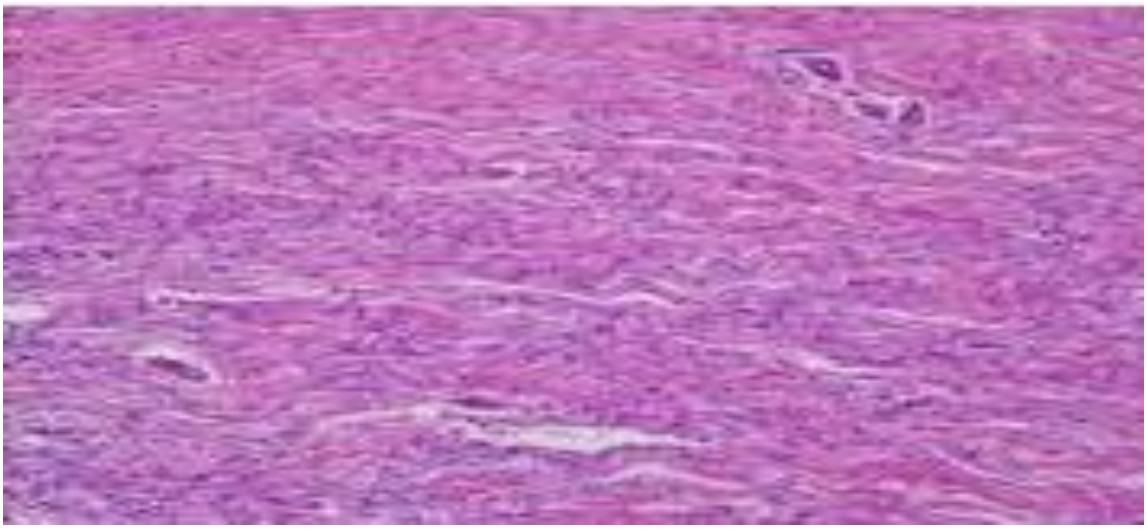
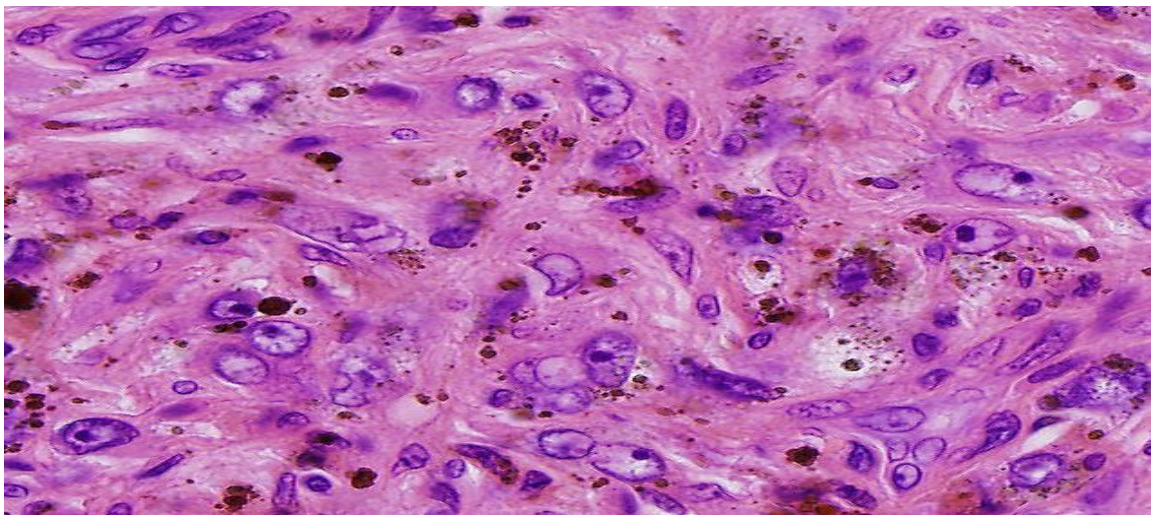
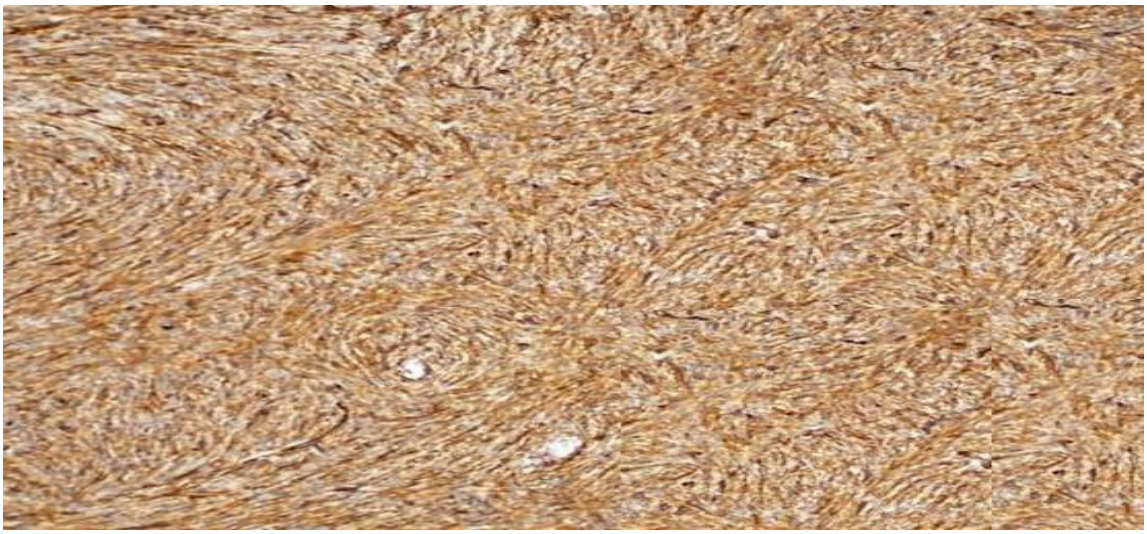


[Fig-4: broad microscopic view of the lesion sample]



[Fig-5: spindle form cells at lesion]



[Fig -6: Immunohistochemical study by avidin-biotin peroxidase technique used to show positivity of antibody against CD34]

III. DISCUSSION

Initially it was known as "*storiform* neurofibroma", this type of dermatofibrosarcoma protuberans contains melanic pigment abundently . It usually occurs after 20 years of age, but it is also found in the children, and called as congenital Bednar tumor. Due to its rareness, the identification of its behavior, treatment and metastasis is based on characteristics of closely related dermatofibrosarcoma protuberans. Although the treatment is surgery, with a safety measure, recurrences may be expected.

The lesions shows a slow growth rates, takes months and years of time.in this case it is found on lateral side of forehead. Lesions are exophytic, nodular, multilobular and of firm consistence. growth invde into the dermis,reachs the subcutaneous layer, musculature.

Due to rare availability, it is difficult to identify Bednar tumor. Commonly suggested diagnosis of melanoma lesion, so histopathologic and immunohistochemical exams are necessary for the correct diagnosis.

The cellular population exhibits elongated, fine elongations, partially or completely enclosed in basal membrane. Another cell population consists of dendritic cells containing melanosomes and premelanosomes.

In immunohistochemical studies most of the tumor cells shows a positive reaction to CD 34 and vimentin.

The treatment is Mohs' micrographic surgery. In the case reported here, the correct diagnosis was not reached initially, but determined later by histopathologic and immunohistochemical examination.

IV. CONCLUSION

From the above study it can be summed up that Bednar tumor, which is very rare type of dermatofibrosarcoma form is mainly associated with pigmented component in this case. As we studied this is a disease with very poor diagnosis process but due to immunohistochemical and histopathological study it can provide a confirmation of the tumor. And only micrographic surgery will be the answer for the treatment of the disease

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