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Case Report

Orthopaedics Section

Isolated Plexiform Neurofibroma of Arm with Unusual Presentation - A Rare Case Report

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ABSTRACT

Plexiform neurofibroma (PNF) arises as a diffuse mass from nerve trunk and leads to overgrowth of cutis and subcutis structure. This is a case report of 20-year-old male, presented to our hospital with a giant ulcerated swelling over his left arm. Differential diagnosis of sarcoma, neurofibroma, hemangioma and angiolipoma was made but biopsy confirmed the diagnosis of plexiform neurofibroma. Isolated PNF with ulceration of overlying skin over arm is a rare presentation and here we are presenting it as a perusal of rare entity.

Keywords: Arm, Neurofibromatosis, Plexiform neurofibroma

CASE REPORT

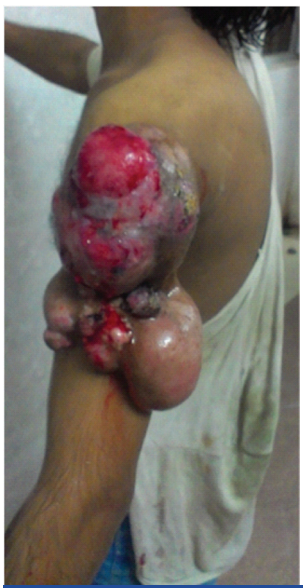
This is a case report of 20-year-old male, presented to our hospital with a giant ulcerated swelling over his left arm. On history taking he revealed that he had noticed this swelling about 12 y back. Initially it was painless and gradually progressive in nature and attained this size in 12 y. For this swelling he did not seek any medical advice and he became used to for it, in his day to day life. Now since six months he developed multiple bleeding points over swelling which were painful. There were no any other associated features, such as other swelling in body or significant family history.

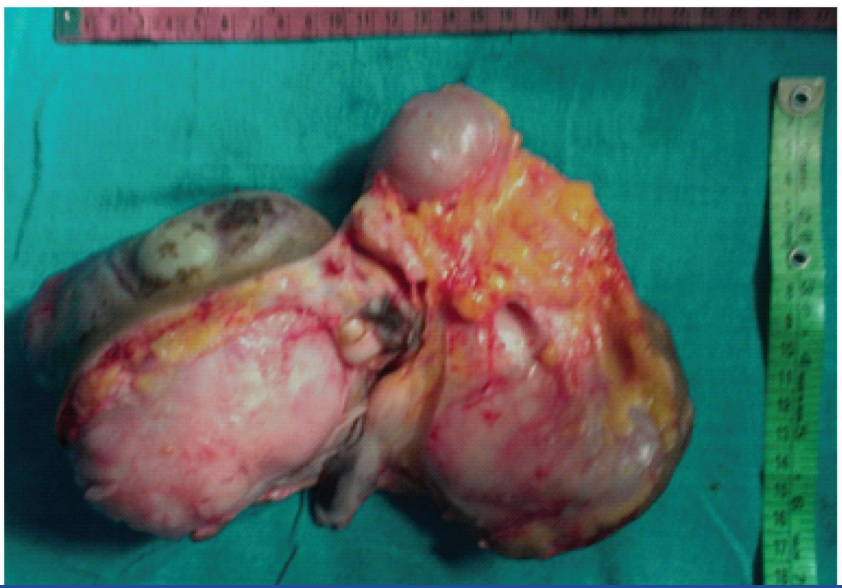
On examination there was huge ulcerated swelling, with firm consistency over his left arm. It was like cauliflower in size and shape, showing venous dilatation over it and occupied the upper two third of left arm from anterolateral to posterolateral aspect [Table/Fig-1]. Differential diagnosis of sarcoma, neurofibroma, hemangioma and angiolipoma was made by clinical examination. X-ray and ultrasonographic modalities sought. In X-ray, humerus was normal and sonography showed echoic mass arising from superficial plain of arm.

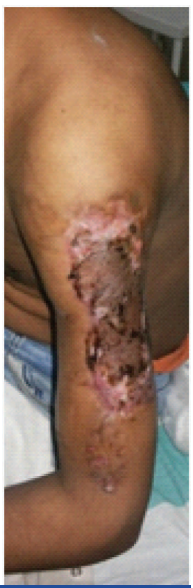
With informed consent, patient planned for excisional biopsy. Since overlying skin was unhealthy, and local skin flap was not available so the swelling was removed en mass from the base including subcutaneous tissue. Specimen weighed of 4500 gm and 22 cm and 17 cm in dimension [Table/Fig-2]. Total blood was about 300 ml and no any intraoperative complication noticed. Since the lesion was bigger in size and due to ulceration infection was expected so cellulitis, lymphedema and delayed healing were conceded as a complications, so the surgical wound left for 3 to 4 days for dressing and thereafter wound covered with skin grafting [Table/Fig-3]. Cut section of swelling showed myxoid like material in between of fatty structure. Part of it removed and sent for biopsy. And the biopsy confirmed the diagnosis of plexiform neurofibroma, without any evidence of malignant changes. His recuperation remained uneventful, and after two weeks patient got discharged, and resumed his work. After one year of follow-up patient is fit and did not show any recurrence.

DISCUSSION

Plexiform neurofibroma (PNF) arises as a diffuse mass from nerve trunk and leads to overgrowth of cutis and subcutis structure and gives a characteristic appearance of “Bag of worm” appearance [1]. Pain, aesthetic issues and neurological involvements are known potential complications of PNF over the certain areas of body, and in a series, 10% of PNE have been undergone into malignant







[Table/Fig-1]: Showing the extension of the ulcerated swelling over the left arm **[Table/Fig-2]:** The excised en mass swelling **[Table/Fig-3]:** The surgical wound managed by superficial skin grafting

changes [2,3]. While it is a matter of debate amongst literature, that what is proper time and proper extent of PNF resection. Some literature suggests that removal of PNF in early stage can restricts the extent of local involvement. But on the contrary Gutmann et al., stated that since complete removal is not feasible sometimes, so in asymptomatic children resection is not justified due to chances of significant regrowth potential of remnant tumor [4,5].

High recurrence tendency of PNF is more with its invasive subtype. In comparison to complete removal (20%), incomplete resection of PNF has (44%) more chances of recurrence. Bounteous bleeding and fragile consistency devotes it to bleed profusely, and makes its complete resection incomplete sometime. Largest series of surgically managed PNF by Needle et al., (in 10 y of follow up) found 54% recurrence rate, and interestingly it reoccurred more in the head and neck area [6]. Extensive surgical approach must be contemplated because inevitable functional deficits have occurred sometimes [7]. That is why, Friedrich et al., in his literature recommended complete removal of small PNF as an inhibitory strategy for later functional deficits [8]. Excision of PNF is a tough task due to their bigger size and intrusion into tissue boundaries.

CONCLUSION

Fortunately, we did not face any problem in this case and here we are sharing our experience with this case of an unusual presentation, which was treated successfully with surgical excision. Isolated PNF with ulceration of overlying skin over arm is a rare presentation and here we are presenting it as a perusal of rare entity. Patient had excellent recovery after resection and no recurrence after one year of follow up till date.

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