Case Report

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Bednar's tumor at right shoulder in an adult male: a case report of a rare entity

Bimarsh Acharya, MBBS[a,*], Dev K. Yadav, MBBS[a], Ankita Chetry, MBBS[a], Pooja Chhetri, MBBS[b], Priya Chhetri, MBBS[c], Divyani Shah, MBBS[d]

Introduction and importance: Bednar tumor, a rare variant of dermatofibrosarcoma protuberans is a tumor with low malignant potential. Owing to limited studies, the pathogenesis and risk factor of this tumor are not clearly understood. The tumor extends from the epidermis to the dermal layers and even affects the surrounding bone. Here, the authors present a case of a 48-year-old male with a Bednar tumor, which was successfully managed by wide local excision and flap repair.

Case presentation: Our patient presented to the hospital after the reappearance of mass, 6 years after the excision of the previously misdiagnosed lipoma. Vitals and systematic examination were normal. Local examination revealed an oval-shaped mass with a glistening surface on the right shoulder. The patient underwent wide excision and skin flap surgery for the treatment, and the diagnosis was confirmed through histopathological examination and immunohistochemistry for the CD34 marker.

Clinical discussion: Bednar tumor is an infrequent skin tumor linked to genetic anomalies and is one of the rare variants [(< 0.1%) of skin tumors]. Diagnosis can be done by histopathological examination and CD34 marker positivity via immunohistochemistry. This tumor is mistaken for lipoma, leading to recurrent growth postexcision. The tumor demands a broader resection due to the high chances of reoccurrence. Although Mohs micrographic surgery is the ideal approach, its limited availability in resource-constrained settings prompts alternative strategies.

Conclusion: This case highlights the challenges of diagnosis, the rarity of the condition, and the need for vigilant follow-up due to the tumor's propensity for recurrence.

Keywords: case report, dermatofibrosarcoma protuberans, skin graft

Introduction

Bednar tumor, also known as storiform neurofibroma is a rare tumor variant of dermatofibrosarcoma protuberans (DFSP) accounting for only 1–5% of all DFSP[1]. DFSP is recognized as a slow-growing skin tumor with intermediate malignancy potential, which can result in greater local destruction of tissues[2]. Here, we present a rare case of a 48-year-old adult male who was diagnosed with DFSP (Bednar's tumor) and treated with a wide excision and skin flap. The case report has been reported in line with Surgical CAse REport (SCARE) criteria[3].

HIGHLIGHTS

- Bednar's tumor is a rare variant of dermatofibrosarcoma protuberans caused due to genetic anomalies.
- Diagnosis can be done using histopathological examination and immunohistochemistry.
- It can be successfully managed with wide local resection.

Case presentation

A 48-year-old male presented with a complaint of progressive swelling over the right shoulder at the hospital. Ten years back, he had a history of suspected lipoma, which was removed by excision. Six years following the removal, the swelling reappeared at the same site and grew to the size of 7 × 5 cm (Fig. 1) for which the patient visited the hospital for further evaluation and management.

On systemic examination, his vitals were within normal limits and there was no tenderness, discharge, significant weight loss, fever, night sweats, or chills. On local examination, the swelling mass was ~7 × 5 cm present over the right shoulder area with a protruding erythematous tip without discharge and tenderness. The range of movement was within the normal limit with intact distal neurovascular status. An MRI of the right shoulder showed calcific focus along with mild supraspinatus tendinosis and acromioclavicular joint arthrosis. Fine needle aspiration cytology of the lesion was suggestive of a pigmented variant of DFSP. Laboratory investigation showed normal random blood sugar,

[a]KIST Medical College and Teaching Hospital, Lalitpur, [b]Kathmandu Medical College, Sinamangal, Kathmandu, [c]Manipal College of Medical Sciences, Pokhara and [d]B.P. Koirala Institute of Health Sciences, Dharan, Nepal

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*Corresponding author. Address: KIST Medical College, Lalitpur, Nepal. Tel.: +977 9867261599. E-mail: achaaryabimarsh456@gmail.com (B. Acharya).

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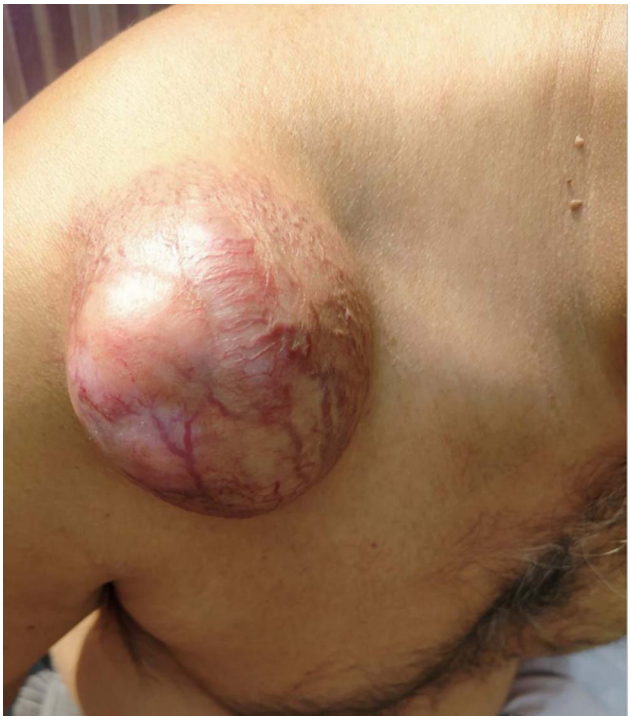

Figure 1. An oval-shaped glistening mass (7 × 5 cm) on the right shoulder over the clavicular region showing prominent veins.

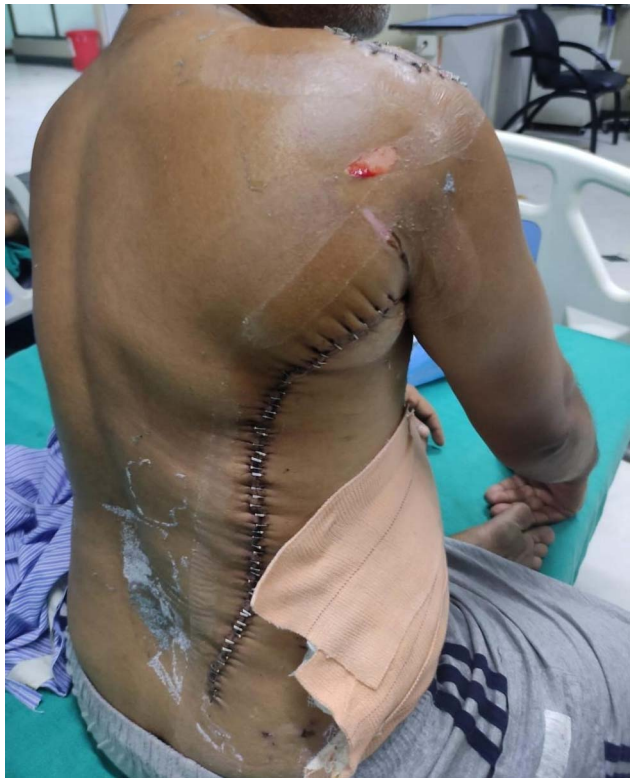

Figure 2. Graft obtained from posterior axilla to loin region, Figure 3: Defect covered with a meshed full-thickness skin graft, Figure 4: Postoperative X-ray showing resected part of the acromioclavicular joint along with stitches and drainage tube.

hemoglobin (13.2 mg/dl) , and packed cell volume (38). Urine routine examination was normal. The patient do not have any prior drug history or family history of similar illness.

Anesthetic consultation was done a day before surgery. A team of orthopedic surgeon and residents performed the surgery. The patient was placed on right lateral decubitus position. Following the marking for incision with a peripheral margin of 7 cm, wide excision of the mass was done along with resection involving part of the acromioclavicular joint to acquire a negative margin. Then the lesion was covered with a meshed full-thickness skin graft along with the stitches around margin of incision. The flap was obtained through a vertical curvilinear excision from the posterior axilla to the loin region (Fig. 2). The specimen was sent to the lab, which tested positive for CD34 marker.

The histopathological examination (HPE) revealed tumors extending from the upper dermis, beneath the centrally atrophic epidermis having focal dense collections of neutrophils infiltrating into subcutaneous fat. There was the storiform and fascicular arrangement of the spindle cells within the compact tumor. Dendritic cells containing melanin pigment were seen scattered in between the spindled cells toward the upper dermis with atypical mitosis (4/10 HPF). This was further supported by immunohistochemistry (IHC) where 76% of cells were immunoreactive with the CD34 marker hence confirming the diagnosis of DFSP of Bednar variant margins were free of the tumor.

The patient was discharged after 9 days of hospital treatment with medication of antidepressants, vitamins, zinc tabs, pantoprazole, and painkillers. The suture removal was done after 12 days. The patient was satisfied with the treatment and was advised for close follow-ups as there are chances of recurrences.

Discussion

DFSP is a rare skin tumor that occurs due to translocation t (17;22)(q22;q13) or chromosome trisomies in the chromosome regions 17q22 and 22q13 due to fusion of alpha chain type 1 of the collagen gene with the platelet-derived growth factor (PDGFB) beta gene resulting in up-regulation of the PDGFB gene[4]. The histological subtypes of DFSP are Bednar tumor, giant cell fibroblastoma, atrophic, sclerosing, granular, and fascicular[5].

Bednar tumor is a rare variant (< 0.1% of skin tumors) of DFSP with tumor cells expressing CD34 + , MART-1 + , and negative S-100 protein[6]. These tumors reportedly occur on trunks, shoulder, upper and lower proximal extremities and rarely in the head and neck[7], which in our case was at the shoulder region. The risk factors of DFSP (Bednar's tumor) are unclear; however, some studies have included previous injuries like burns, pre-existing scars, and tattoos as risk factors, while gender predominance is not clear[8–10]. The prevalence, associated risk factors, and management of Bednar tumor are very rarely reported, which makes this case report valuable.

The diagnosis of Bednar tumor (DFSP) is established by HPE, as is done in our case. Under microscopy, Bednar tumor appears as a matting-like (storiform neurofibroma) structure found only at the center of the tumor, which consists of a group of spindle-shaped tumor cells giving the appearance of little stars or bilateral brushes. The histological findings in our case are similar to most of the reported cases. Occasionally, it can be confused with

dermatofibroma (DF)[5,11]. Several studies suggest the majority of DFSP cases have CD34+, Factor XIIIa-, and CD68-immunophenotypes[12–14]. Our case also underwent IHC, which was positive for the CD34 marker, which provided extra support during diagnosis.

Cases had been reported where Bednar tumor (DFSP) had been misdiagnosed as lipoma initially, and recurrence of the same mass after excision[15–17]. Similarly, in our case, the mass, which was initially misdiagnosed and treated as lipoma, recurred and then later diagnosed as Bednar tumor. For this, wide local excision was done, the margin of which was comparatively more in our case than the recommended standard margin for similar cases[3]. Standard surgical treatment for DFSP is Mohs micrographic surgery also known as staged wide excision done with histopathological sectioning of the tumor and delayed reconstruction for complete circumferential peripheral and deep margin assessment is done[5,9,11]. The recurrence rate after Mohs surgery is low but in low-resource setting countries like Nepal, such procedures are not done routinely due to high cost and less availability of treatment modalities[18].

Conclusion

Bednar tumor is a rare malignant entity with lesser life-threatening complications; however, it can be a cosmetic nuisance. The chances of misdiagnosis are relatively higher in low-resource setting countries. The diagnosis of Bednar tumor can be done by HPE and IHC (CD34+). Wide local excision with extra margin can be done if Mohs micrographic surgery is unavailable. The tumor has a high recurrence rate even after surgery, for which follow-ups should be done routinely.

Ethical approval

This is a case report therefore, it did not require ethical approval from the ethics committee.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

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All data are provided within this review and data within original published papers noted in this review.

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