DOI: 10.7860/JCDR/2022/50393.16358

Case Report

Surgery Section

A Case of Bednar Tumour in the Sacral Region- A Rare Pigmented Variant of Dermatofibrosarcoma Protuberans

JAISTRI JAISHANKAR[1], R ANANTHARAMAKRISHNAN[2], K SENTHIL KUMAR[3], VIJAYASHREE RAGHAVAN[4]

ABSTRACT

Dermatofibrosarcoma Protuberans (DFSP) is a relatively rare soft tissue neoplasm of intermediate malignancy. Metastasis of this tumour seldom occurs. The DFSP is a locally aggressive tumour with a high recurrence rate. The DFSP accounts for less than 0.1% of all malignant neoplasms and approximately 1% of all soft tissue sarcomas. Bednar tumour is an uncommon variant of dermatofibrosarcoma protuberans. Also known as pigmented dermatofibrosarcoma protuberans, this tumour is a rare, aggressive, cutaneous tumour that constitutes 1-5% of all DFSPs of intermediate grade. These tumours can be easily mistaken for fibrosarcoma and melanoma. It is seen in adults with a predisposition to affect the shoulder region, and trunk. Authors hereby report a case of 50-years-old female presented with complaints of recurrent swelling in the lower back for a duration of four months. Pain aggravated on lying in the supine position, swelling was painless otherwise. Patient also had complaints of itching over the swelling with occasional blood stained discharge on self-inflicted trauma. Magnetic Resonance Imaging (MRI) scan and histopathological examination of excised tumour confirmed the diagnosis of bednar tumour. She underwent wide local excision with split skin grafting and was on follow-up for the last two years with no evidence of recurrence. Hence, adequate clearance and thorough and regular follow-up is essential to prevent recurrence.

Keywords: Soft tissue neoplasm, Split skin grafting, Wide local excision

CASE REPORT

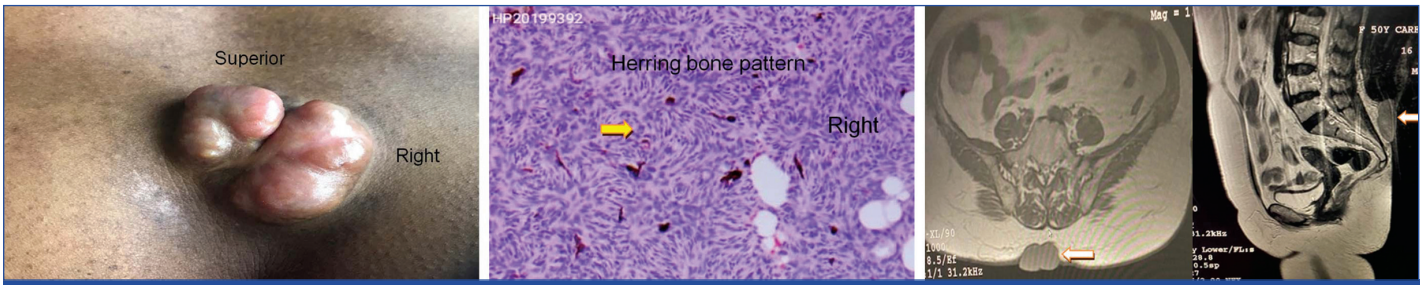
A 50-years-old, postmenopausal, female presented to the General Surgery Outpatient Department (OPD) with complaints of recurrent swelling in the lower back for a duration of four months. Pain aggravated on lying in the supine position with no specific relieving factors, swelling was painless otherwise. Patient also had complaints of itching over the swelling with occasional blood-stained discharge on self-inflicted trauma. There was no history of changes in the surrounding skin, fever, trauma, congenital malformations, bladder or faecal incontinence, headache, neck rigidity, weakness of limbs, difficultly in walking, similar swellings elsewhere in the body noticed in the last four months. No history of substance abuse and nil significant family history.

She underwent excision of the swelling elsewhere four years back (records not available) following which she was asymptomatic post procedure for two years. Patient experienced itching at the same site on and off over last two years after which she noticed recurrence of the swelling at the same four months back which was progressive in size and associated with intermittent, non radiating type of pain over the swelling. Pain aggravated only on lying down in the supine position.

On examination of the sacral region, a proliferative, irregular shaped swelling with ill-defined margins approximately 6×4 cm was seen in the sacral region about 2 cm above the gluteal cleft in the midline. Skin over the swelling appears discolored, non pulsatile. No ulcers, dilated veins or sinus over the swelling [Table/Fig-1]. On palpation, no increase in warmth, non tender swelling with a variable consistency, nodular surface, restricted mobility, skin over swelling was not pinchable, no lymph node involvement in the sacral region.

Post admission, blood investigations like complete haemogram, bleeding time, clotting time, random blood sugar, serology were done. All these investigations were within normal limits. A provisional diagnosis of soft tissue sarcoma in the sacral region was made. Trucut biopsy was done which reported findings suggestive of pigmented dermatofibrosarcoma protuberans (bednar tumour) On microscopy, dermis shows ill circumscribed unencapsulated tumour consisting of spindle shaped cells arranged in storiform pattern. Cells show minimal cytological atypia, spindle shaped nucleus with blunt edges, inconspicuous nucleoli and scant cytoplasm. Mitosis was present approximately 4/10 High Power Field (HPF). Collagen trapping seen. Many pigmented cells seen admixed with tumour cells [Table/Fig-2].

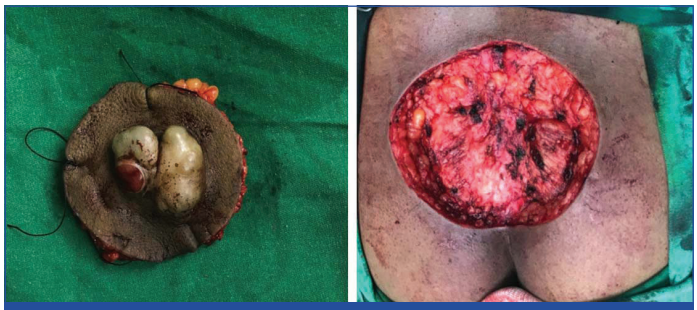
Magnetic Resonance Imaging (MRI) abdomen and pelvis was done which showed exophytic, soft tissue lesion in lumbo-sacral junction level in dorsal aspect with lesion contained within subcutaneous fat plane and without deeper extension [Table/Fig-3].



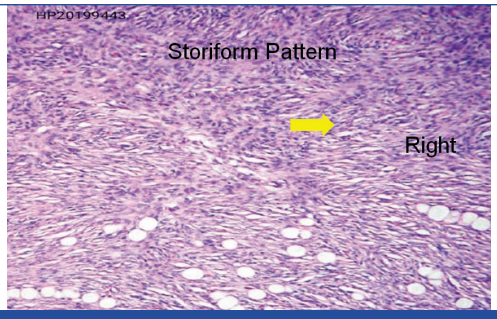
[Table/Fig-1]: Proliferative, irregular shaped swelling with ill-defined margins ~6×4 cm swelling in the sacral region; **[Table/Fig-2]:** Microscopic image of Bednar tumour showing many pigmented cells seen admixed with tumour cells and collagen trapping [H&E stain,10X]; The yellow arrow indicates herring bone pattern of spindle shaped cells. **[Table/Fig-3]:** On MRI of pelvis: T1 weighted image and sagittal section showing the lesion. (Images from left to right)

Patient was then planned for wide local excision with split skin grafting after obtaining fitness for procedure under general anaesthesia.

Patient was put in prone position under general anaesthesia after which parts were painted and draped. Wide local excision of the swelling was done with 3 cm margin all around [Table/Fig-4]. The SSG was taken from the left thigh and placed over the raw area in the sacral region [Table/Fig-5]. Sterile dressing was applied. Clearance was given with the underlying fascia Postoperative period was uneventful. Specimen excised was sent for histopathological examination and was found to be consistent with dermatofibrosarcoma protuberans with all margins being free of tumour [Table/Fig-6].



[Table/Fig-4]: Excised specimen with 3 cm margins all around the tumour.
[Table/Fig-5]: Tumour bed showing complete macroscopic clearance. (Images from left to right)



[Table/Fig-6]: Spindle shaped cells arranged in a storiform pattern [H&E,10X]. The Yellow arrow indicates storiform pattern of spindle shaped cells.

Patient was on a regular follow-up for the last two years and the wound site had healed completely with good uptake of the graft and there were no signs of recurrence.

DISCUSSION

Gunasekaran V et al., suggested that bednar tumour which is a variant of dermatofibrosarcoma protuberans has an intermediate malignancy potential and that wide local excision with marginal clearance is curative. Closure of the defects was given by flap cover [1].

In this case, a split skin graft was done following wide local excision of the tumour and the patient is on regular follow-up till date and there is no evidence of recurrence. Patil P et al., reported a case of DFSP in a 9-year-old boy who underwent excision of the lesion with a 1 cm margin and the patient was followed-up for a period of one and a half years and there no no evidence of recurrence [2].

A rare variant of dermatofibrosarcoma protuberans is bednar tumour which is a pigmented variety described first in 1957 [3]. It is not commonly seen in the general population, and it is considered under intermediate grade of tumour. It is described to have a pigmented irregular surface with its growth extending into the subcutaneous tissue [3]. The striking feature on histology is the presence of spindle shaped cells and dendritic cells which contain melanin.

It is suggested that this tumour is seen to be arising from neuroectodermal cells. The melanin is responsible for the pigmented nature of this tumour [4]. Fibrous histiocytoma, neurofibroma, malignant melanoma, and cellular blue nevus are the differential diagnosis given for bednar tumour [4]. It contain S-100 protein although not always but CD 34 antigen is present in DFSP and Bednar tumour [4]. It has also been stated that patients who present with this tumour are often seen consulting in the dermatology department and Moh's micrographic surgery is one of the treatment options [5].

Nakamura T et al., stated that COL1A1-Platelet Derived Growth Factor subunit B (PDGFB) gene fusion was seen which led to the expression of PDGFB, portraying the tumorigenic mechanism in cases of dermatofibrosarcoma protuberans [6].

Dupree WB et al., stated that it was uncommon to see metastasis in dermatofibrosarcoma protuberans and that complete excision with close follow-up was indicated in this condition as it is a tumour with intermediate potential to transform into malignancy [7].

In this case, wide local excision with clearance which was confirmed by histopathological examination with a split skin graft to cover the defect was done for this patient and the patient is on regular follow-up with no evidence of recurrence till date.

CONCLUSION(S)

A rare variant of dermatofibrosarcoma protuberans, bednar tumour seen in the sacral region with complete excision with no microscopic residue of tumour. It should be differentiated from other conditions which mimic dermatofibrosarcoma protuberans clinically and an adequate clearance with negative tumour margins should be ensured as this tumour has a high rate of recurrence.

REFERENCES

[1] Gunasekaran V, Peters NJ, Samujh R. Congenital giant Bednar tumor in a child: surgical challenges in a young infant: A case report. Ann Pediatr Surg. 2020;16(1):01-04.
[2] Patil P, Tambe S, Nayak C, Ramya C. Dermatofibrosarcoma protuberans in a 9-year-old child. Indian Dermatology Online Journal. 2017;8(3):195.
[3] Amonkar GP, Rupani A, Shah A, Deshpande R. Bednar tumor: An uncommon entity. Dermatopathology. 2016;3(2):36-38.
[4] Kaul R, Kaur N, Dogra SS, Chander B. Variant of dermatofibrosarcoma protuberans: Bednar tumor. Indian journal of dermatology. 2015;60(1):107.
[5] Bednář B. Storiform neurofibromas of the skin, pigmented and nonpigmented. Cancer. 1957;10(2):368-76.
[6] Nakamura T, Ogata H, Katsuyama T. Pigmented dermatofibrosarcoma protuberans. Report of two cases as a variant of dermatofibrosarcoma protuberans with partial neural differentiation. Am J Dermatopathol. 1987;9(1):18-25.
[7] Dupree WB, Langloss JM, Weiss SW. Pigmented dermatofibrosarcoma protuberans (Bednar tumor). A pathologic, ultrastructural, and immunohistochemical study. Am J Surg Pathol. 1985;9(9):630-39.

PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of General Surgery, Chettinad Hospital and Research Institute/Care, Chengalpet, Tamil Nadu, India.
2. Professor, Department of General Surgery, Chettinad Hospital and Research Institute/Care, Chengalpet, Tamil Nadu, India.
3. Professor, Department of General Surgery, Chettinad Hospital and Research Institute/Care, Chengalpet, Tamil Nadu, India.
4. Professor, Department of Pathology, Chettinad Hospital and Research Institute/Care, Chengalpet, Tamil Nadu, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. K Senthil Kumar,
House No-108 (3/2) F2, Sahana Flats, Sayee Nagar, 5th Street, Virugambakkam,
Chennai-600092, Tamil Nadu, India.
E-mail: drsenthilchri@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jul 23, 2021
- Manual Googling: Mar 14, 2022
- iThenticate Software: Mar 22, 2022 (11%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jul 22, 2021**
Date of Peer Review: **Oct 13, 2021**
Date of Acceptance: **Mar 15, 2022**
Date of Publishing: **May 01, 2022**