

Large chordoma of the sacrum

Owen Godkin, Hussam Elkhwad, John McCabe

Department of Trauma and Orthopaedics, Galway University Hospitals, Galway, Ireland

Correspondence to
Dr Owen Godkin,
owen.godkin@gmail.com

Accepted 7 July 2017

DESCRIPTION

A 53-year-old man presented to our service with a 1-month history of subacute bowel obstruction. Clinical examination was unremarkable with normal power and sensation in the lower limbs. This patient had no medical or surgical history. MRI of the pelvis identified a well-defined presacral midline lesion of 9.9 cm diameter, which appeared to arise from the S2 nerve root and below (figure 1A,B). These nerve roots control anal sphincter tone and prohibit faecal incontinence. Whole spine MRI and CT-thorax abdomen pelvis CT-TAP out ruled metastasis. A biopsy confirmed diagnosis of chordoma. This patient was discussed by a multidisciplinary team consisting of orthopaedics, colorectal, vascular and plastic surgeons. Anterior mobilisation of the lesion, with ileostomy due to expected loss of anal tone and supporting vessel ligation, was undertaken. A wide sacral *en-masse* resection at the level of S1 disc and inferior nerve roots, incorporating partial gluteus maximus and piriformis bilaterally, was achieved by a posterior approach with a latissimus dorsi flap for reconstruction (figure 2A,B,C). Histopathology confirmed clear margins.

Chordomas are rare slow-growing malignant tumours of embryonic notochord remnants. They comprise 1%–4% of all primary bone tumours.¹ Most common locations include the base of skull and sacrococcygeal regions. Sacrococcygeal chordomas can affect bowel, urinary and vascular structures resulting in obstruction. Diagnosis includes plain radiography, MRI and CT-TAP to identify metastasis. Definitive diagnosis is achieved with a tissue biopsy. Differential diagnosis includes chondrosarcoma and tuberculosis. Treatment by surgery is a wide-margin *en-masse* resection. Poor prognosis has been identified with a large tumour size

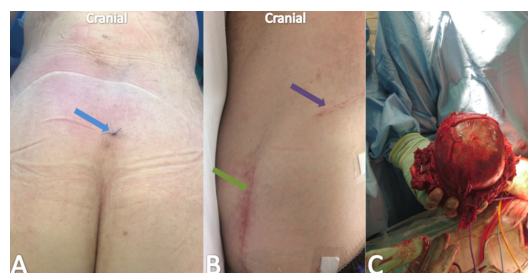


Figure 2 Topographical view of patient's lumbar spine; preoperative (A) with suture identifying the biopsy site (blue arrow) and 6 weeks postoperative (B) with sacral (green arrow) and latissimus dorsi (purple arrow) wounds. Chordoma lesion (C) and sacrum excised, *en-masse*.

and incomplete marginal clearance.² Clinical trials with immunotherapy, supported by the chordoma foundation, are currently underway for advanced metastatic disease.³

Learning points

- ▶ Chordomas are rare slow-growing malignant tumours of embryonic notochord remnants.
- ▶ Symptoms vary depending on compressive features.
- ▶ Surgical resection is the primary goal with poor prognosis related to size and age of onset.

Contributors OG and HE: image and data gathering. OG and HE: writing of manuscript. JMC: clinical supervisor and treated the patient.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Hsieh PC, Xu R, Sciubba DM, *et al.* Long-term clinical outcomes following en bloc resections for sacral chordomas and chondrosarcomas: a series of twenty consecutive patients. *Spine* 2009;34:2233.
- 2 Bergh P, Kindblom LG, Gunterberg B, *et al.* Prognostic factors in chordoma of the sacrum and mobile spine: a study of 39 patients. *Cancer* 2000;88:2122–34.
- 3 Chordoma Foundation. Now open: immunotherapy research study designed specifically for chordoma patients. <https://www.chordomafoundation.org/latest-updates/nivolumab-chordoma-clinical-trial-now-open/> (accessed 31 May 2017).

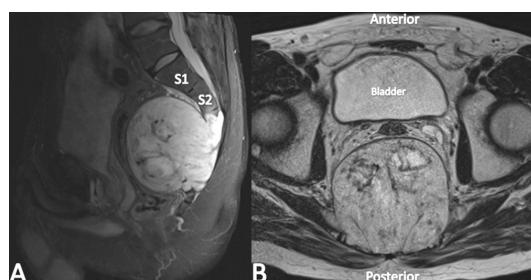


Figure 1 A sagittal (A) and axial (B) MRI of a well-defined midline 9.9 x 9 cm arising from the sacral spine.



To cite: Godkin O, Elkhwad H, McCabe J. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221293

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow