RCS ADVANCING SURGICAL STANDARDS

ONLINE CASE REPORT

Ann R Coll Surg Engl 2014; **96:** e8–e11
doi 10.1308/003588414X13946184902686

Hand metastasis from a sacral chordoma

M Emori[1], M Kaya[1], S Sugita[1], T Soma[2], M Sasaki[1], T Yamashita[1]

[1]Sapporo Medical University, Japan
[2]Hokkaido Cancer Center, Sapporo, Japan

ABSTRACT

Chordomas are rare, low grade, malignant tumours derived from the ectopic remnants of the notochord that line the axial skeleton. They are characterised by their slow growth, long disease course and propensity for local relapse. Furthermore, up to 40% of non-cranial chordomas metastasise. We describe the first reported case of a hand metastasis arising from a conventional sacral chordoma after carbon ion radiotherapy. The common occurrence of distant metastasis with chordomas makes it important to perform a systemic examination, in part because their resection might improve patient prognosis.

KEYWORDS

Chordoma – Hand metastasis – Carbon ion radiotherapy

Accepted 14 October 2014; published online XXX

CORRESPONDENCE TO

Makoto Emori, E: emrmkt@yahoo.co.jp

Chordomas are very rare malignant bone tumours that develop from the remnants of the primitive notochord and arise in the sacrum, spine or base of the skull. There is currently no standard systemic therapy and surgery is the most common treatment option. The achievement of negative surgical margins is associated favourably with the rate of local relapse and survival.[1] However, complete tumour resection is sometimes difficult to achieve if the tumour volume is large and/or the tumour invades the upper levels of the sacrum. Furthermore, complete resection can potentially result in functional deficit owing to the impairment of major nerves, which will in turn adversely affect the patient's quality of life. There is therefore a growing acceptance of carbon ion radiotherapy (CIRT) as an alternative to surgery for unresectable sacral chordomas. CIRT is associated with a higher local control rate as well as better preservation of urinary and anorectal function than surgery.[2]

We describe what we believe to be the first reported case of a hand metastasis originating from a chordoma after CIRT. Our case illustrates the importance of a systemic examination to detect distant metastases of sacral chordoma.

Case History

A 71-year-old woman presented with swelling and pain in her buttock, which she had first noticed 3 months previously and which had worsened steadily. Plain radiography of the sacrum showed a destructive and radiolucent lesion of a soft tissue mass. Magnetic resonance imaging (MRI) showed that the lesion was homogeneously isointense to skeletal muscle on a T1 weighted image, and heterogeneously hypointense and hyperintense on a T2 weighted image. Its apparent size was 11cm × 6.4cm × 6.8cm (Fig 1). Open biopsy was performed, and the pathological diagnosis was conventional chordoma, based on the results of imaging studies together with those of histologic analysis of the biopsy specimen. The tumour had a lobulated architecture composed of epithelioid cells with vacuolated cytoplasm that contained glycogen in a mucoid matrix and tumour cells

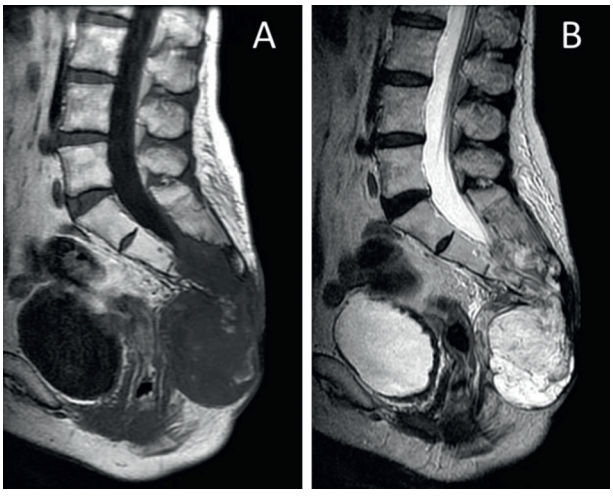

Figure 1 Sagittal magnetic resonance imaging showing that the lesion was homogeneously isointense to skeletal muscle on T1 weighted imaging (A), and heterogeneously hypointense and hyperintense on T2 weighted imaging (B)

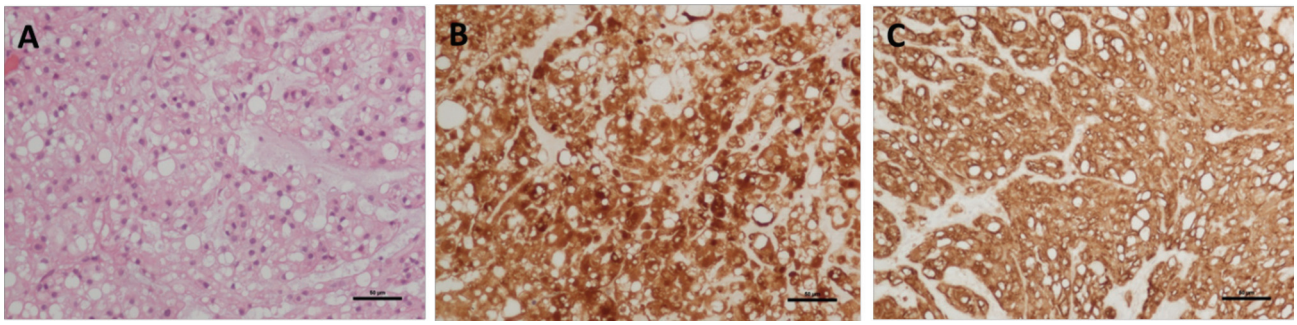

Figure 2 Histological analysis of the primary tumour: haematoxylin and eosin staining (A), and immunohistochemical staining for S100 (B) and AE1/3 (C) proteins

were positive for S100 and AE1/3 protein immunohistochemistry (Fig 2).

The tumour extended as far the S2 region. As a result, a procedure that was sufficiently extensive to achieve a negative surgical margin carried a significant risk of nerve damage and functional impairment. After consultation with the patient, it was decided that treatment should consist of CIRT rather than surgery.

Eighteen months after treatment, the patient presented with swelling of the right thumb and, on examination, a firm mass was found in the thenar area (Fig 3). Plain radiography of the left hand showed a soft tissue mass with destruction of the first metacarpal bone (Fig 4) and MRI revealed a tumour located in the thenar region with invasion to the first metacarpal bone.

Measuring 18mm × 28mm × 35mm, the tumour showed homogeneous hypointensity to skeletal muscle on T1 weighted imaging with gadolinium enhancement whereas it

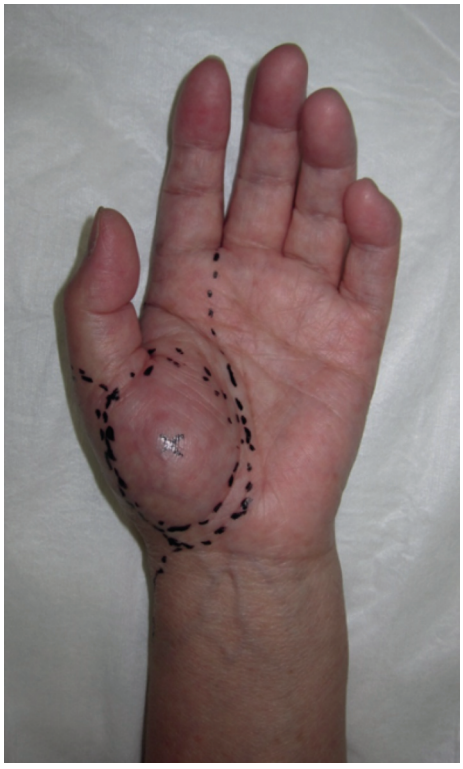

Figure 3 The tumour was elastic and soft with a relatively clear border, located in the thenar region.

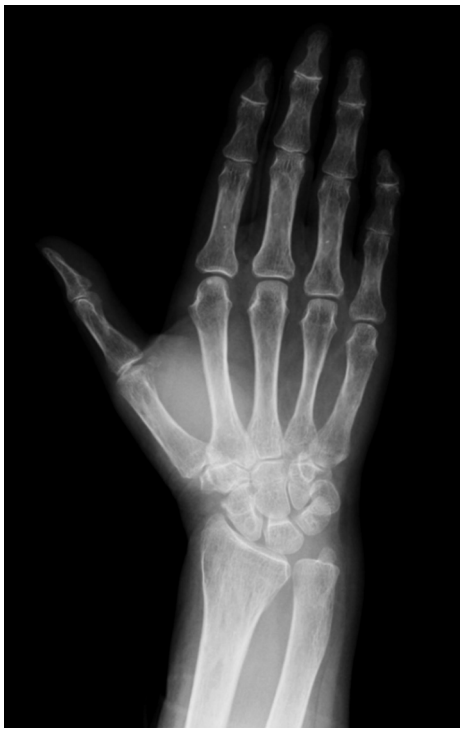

Figure 4 Preoperative plain radiography of the left hand showing soft tissue mass and erosion of the first metacarpal bone

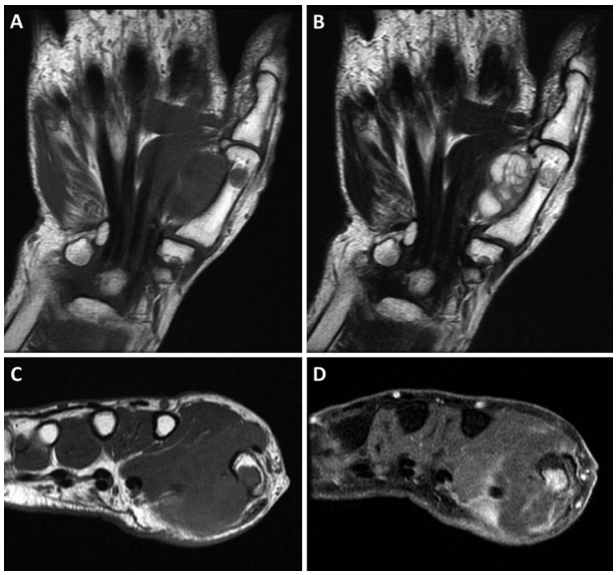

Figure 5 Magnetic resonance imaging showing a soft tissue mass lesion arising in the thenar region eroding the first metacarpal bone: coronal T1 weighted image (A), coronal T2 weighted image (B), axial T1 weighted image (C) and gadolinium enhanced image (D)

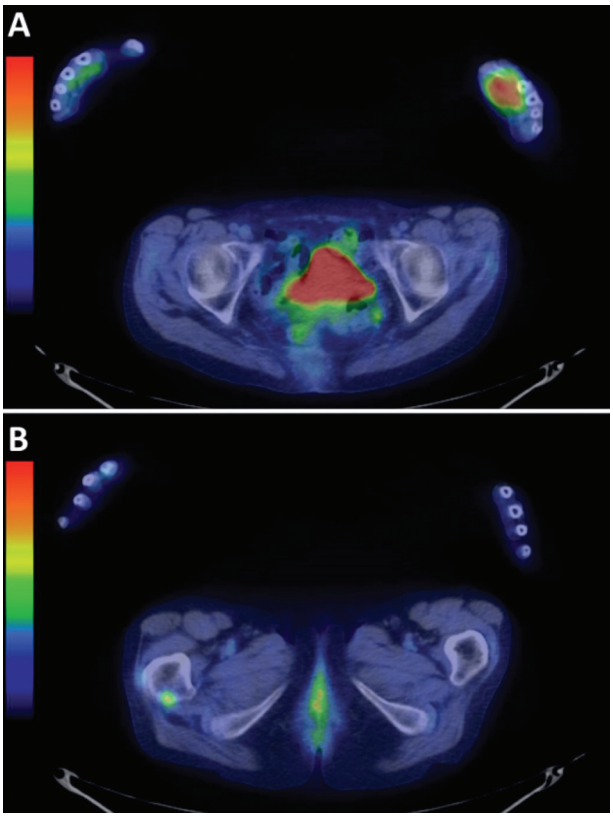

Figure 6 Positron emission tomography showing increased uptake of fludeoxyglucose in the thenar (A) and right proximal femur region (B)

was heterogeneously isointense and hyperintense on T2 weighted imaging (Fig 5). Fludeoxyglucose positron emission tomography (FDG PET) was performed to screen for metastases. Increased uptake of fludeoxyglucose was observed in the thenar, right proximal femur and left pulmonary lower lobe regions (Fig 6). A needle biopsy was performed to confirm whether the hand lesion was a metastatic tumour, which revealed it to have the same pathological features as the primary tumour (Fig 7).

On the basis of these findings, the tumour was diagnosed as a metastatic chordoma. In order to achieve negative resection margins, tumour excision comprised double first and second ray amputations involving the trapezium, trapezoid and navicular bones (Fig 8). The procedure also involved wide resection of the proximal femur with reconstruction using a Kotz prosthesis and partial resection of the left lower lobe. At a follow-up screening nine months later, there was no apparent recurrence or metastasis and the patient was able to walk with the aid of a cane.

Discussion

CIRT has been used increasingly as an alternative to surgery for the treatment of medically unresectable sacral chordomas and it has resulted in good local control.[3] The local control rate has been reported to be approximately 60–80% in cases where the tumour was totally excised[4,5] while the five-year local control rate for CIRT treated cases is reported as 88%.[3] The rate of metastasis for chordomas

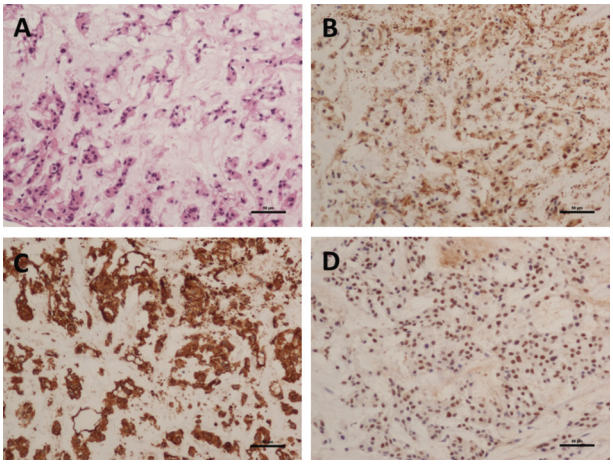

Figure 7 Histological analysis of the metastatic tumour: haematoxylin and eosin staining (A), and immunohistochemical staining for S100 (B), AE1/3 (C) and Brachyury (D) proteins

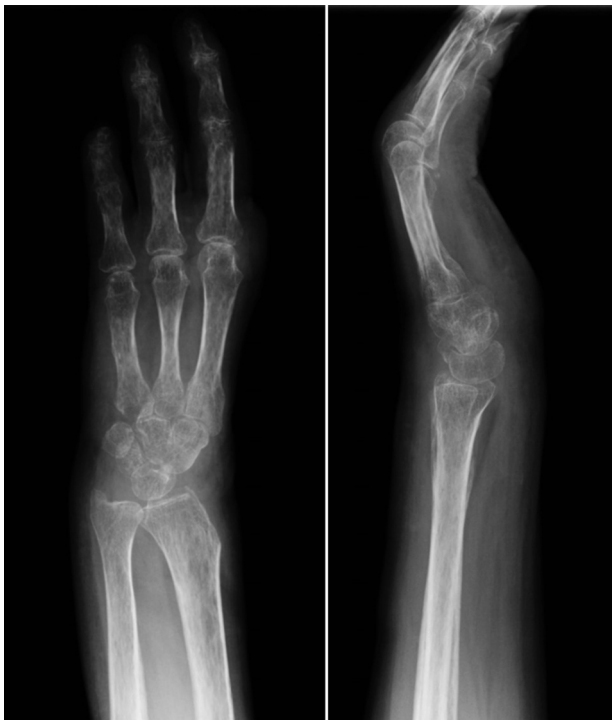

Figure 8 Plain radiography obtained at the latest follow-up visit

after CIRT has been reported as 24.7%,[6] which was similar to that achieved in some relatively recent studies.[7] Consequently, it remains unclear as to how effective CIRT is at preventing metastasis in chordomas.

Previous studies have suggested that the incidence of metastases in this malignancy ranges from 5% to 43%.[6] The most common site of metastasis is the bone, followed by the lung and liver. Among bone metastases, the iliac bone is the most often involved.[6] Regardless of the site of metastasis, patient survival seems to be affected more by local progression than by the metastasis itself.[8] It is therefore unknown whether the resection of distant metastatic lesions affects the outcome of this disease.[9]

We have described what we believe to be the first reported case of hand metastasis arising from a conventional chordoma after CIRT. Metastatic spread of cancer to the hand is also rare, representing only 0.1% of all cancer metastases.[10] Reported examples include lung cancer and renal cell carcinoma metastasising to the hand, and, in most cases, patients die of the disease within six months.[11]

Conclusions

Chordomas are slow growing tumours and so resection of distant metastasis might improve the prognosis. Accordingly, we suggest that it is important to perform a systemic examination, for example using PET or whole body MRI.

References

1. Baratti D, Gronchi A, Pennacchioli E *et al.* Chordoma: natural history and results in 28 patients treated at a single institution. *Ann Surg Oncol* 2003; **10**: 291–296.
2. Nishida Y, Kamada T, Imai R *et al.* Clinical outcome of sacral chordoma with carbon ion radiotherapy compared with surgery. *Int J Radiat Oncol Biol Phys* 2011; **79**: 110–116.
3. Imai R, Kamada T, Sugahara S *et al.* Carbon ion radiotherapy for sacral chordoma. *Br J Radiol* 2011; **84**: S48–S54.
4. Ozaki T, Hillmann A, Winkelmann W. Surgical treatment of sacrococcygeal chordoma. *J Surg Oncol* 1997; **64**: 274–279.
5. Yonemoto T, Tatezaki S, Takenouchi T *et al.* The surgical management of sacrococcygeal chordoma. *Cancer* 1999; **85**: 878–883.
6. Kishimoto R, Omatsu T, Hasegawa A *et al.* Imaging characteristics of metastatic chordoma. *Jpn J Radiol* 2012; **30**: 509–516.
7. Stacchiotti S, Casali PG, Lo Vullo S *et al.* Chordoma of the mobile spine and sacrum: a retrospective analysis of a series of patients surgically treated at two referral centers. *Ann Surg Oncol* 2010; **17**: 211–219.
8. Boriani S, Chevalley F, Weinstein JN *et al.* Chordoma of the spine above the sacrum. Treatment and outcome in 21 cases. *Spine* 1996; **21**: 1569–1577.
9. Chambers PW, Schwinn CP. Chordoma. A clinicopathologic study of metastasis. *Am J Clin Pathol* 1979; **72**: 765–776.
10. Kerin R. The hand in metastatic disease. *J Hand Surg Am* 1987; **12**: 77–83.
11. Wu KK, Guise ER. Metastatic tumors of the hand: a report of six cases. *J Hand Surg Am* 1978; **3**: 271–276.