Clinics in Diagnostic Imaging (65)

M Y S Soo

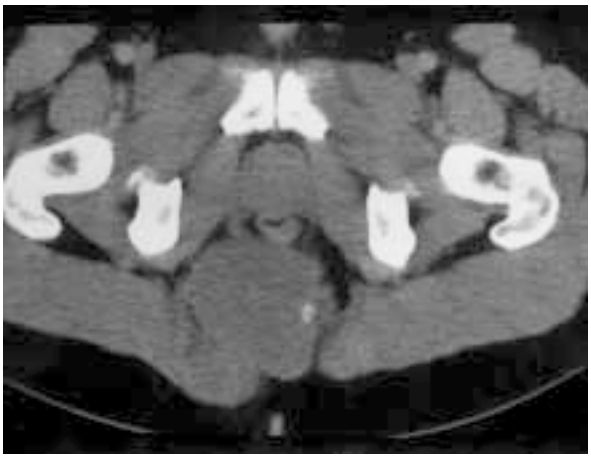

Fig. 1a Enhanced axial CT taken through the level of the pelvic floor.

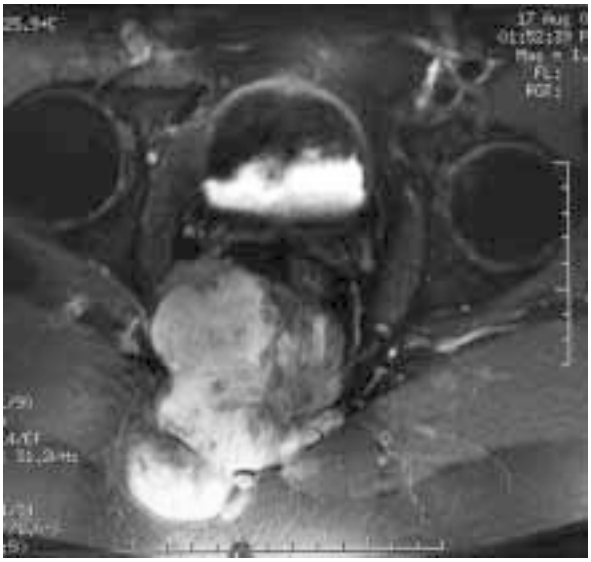

Fig. 1b Enhanced axial T1-weighted MR image taken 3 cm cranial to Fig. 1a.

CASE PRESENTATION

A 41-year-old man of Maori extraction gave a history of progressive pain in the gluteal region for the past five months. The pain was made worse on sitting. Over the same period, he noticed a tender lump in the right buttock. There was no relevant information in the past history. Physical examination revealed a hard lump of about 5 cm in diameter in the subcutaneous and deep tissues of the right buttock. Rectal examination confirmed the location of the lesion in the right ischiorectal fossa. The rectal wall was intact. Computed tomography (CT) and magnetic resonance (MR) imaging were performed (Figs. 1a-b). What do the images show? What is the diagnosis?

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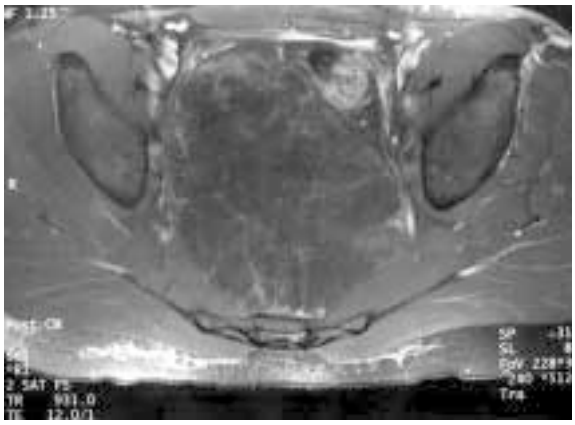

Fig. 2 19-year-old Caucasian man who presented with perineal pain and constipation for four months. Enhanced axial fat suppressed T1-weighted MR image taken through this large pelvic tumour shows only patchy enhancement. Note the numerous septations contrasted against the dark greyish tone of the tumour tissue. The pyriformis muscles and the 4th sacral segment are intact. In this case, the chordoma arose from the coccyx. There is enhancement of the more superficial layer of the right gluteus maximus muscle.

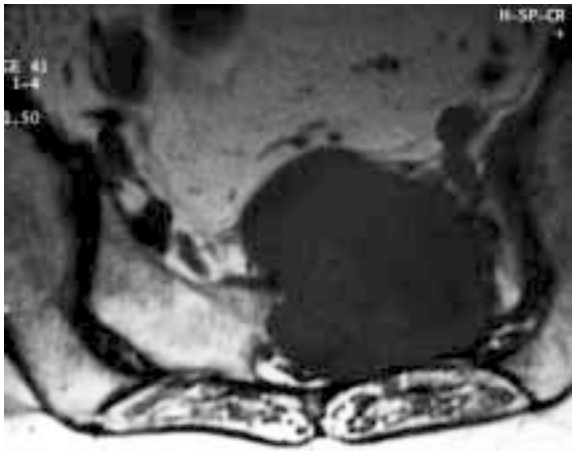

Fig. 3a Axial T1-weighted MR image of a 78-year-old Caucasian woman suffering from two years of back pain. A 7 cm diameter well-defined homogeneously hypointense lesion is seen arising from the sacral canal and left S3 body. A greater portion of the tumour occupies the sacrum and only a proportionately smaller component extends anteriorly to the presacral space.

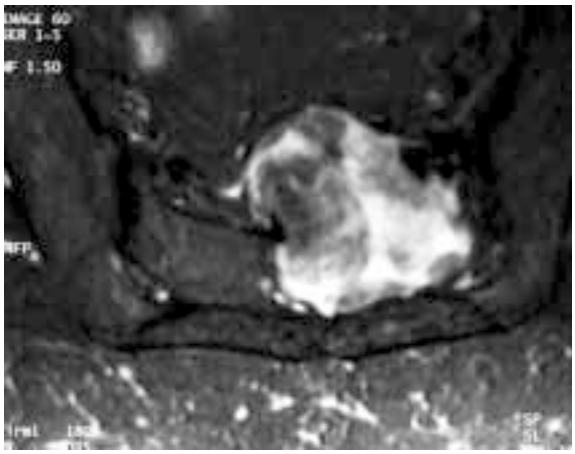

Fig. 3b Axial STIR MR image of the same patient as Fig. 3a. Nodular foci of hypointensity are seen within a predominantly hyperintense lesion. No internal septations are visible. An open biopsy was performed. Histopathology was neurilemmoma.

IMAGING INTERPRETATION

Enhanced axial CT scan (Fig. 1a) shows a large 5 cm diameter soft tissue mass with well-defined margins arising from the sacrococcygeal region. It is faintly calcified, invades the right gluteus maximus muscle, and encroaches on the fat plane of the right ischiorectal fossa. The invaded portion of the right gluteal muscle is swollen. No obvious contrast enhancement is noted. The rectal wall and the obturator internus muscles are intact.

On the enhanced axial T1-weighted MR image, (Fig. 1b) there is a marked degree of contrast enhancement. Numerous linear to nodular low signal septations are seen within the lesion. Lesional invasion of the right gluteus maximus and obturator internus muscles is obvious. Extension into the medial portion of the left gluteus maximus muscle is also evident. In keeping with the CT appearances, the lesion is shown to be centred on and possibly arising from the lower sacrococcygeal region.

DIAGNOSIS

Sacrococcygeal chordoma.

CLINICAL COURSE

The patient underwent surgery to remove the tumour. A combined lower abdomino-sacral approach was used. An anterior midline incision was made to ligate both superior gluteal arteries. This was followed by a posterior midline approach over the sacrum to remove the chordoma which arose from the S4 segment. Complete excision was achieved with minimal blood loss. Histopathological diagnosis was that of a well-differentiated chordoma. The patient made a good recovery and was discharged from hospital 10 days later.

DISCUSSION

Chordomas are rare malignant bone tumours that originate from the remnants of the primitive notochord. They are thus characteristically located at the skull base and vertebral column. They account for about 1% of all primary bone tumours[1]. Sites of predilection are the sacrococcygeal regions (50%) and spheno-occipital synchondrosis (35%). The mobile spinal column is involved in 15% of cases, most commonly in the upper cervical spine[2]. With spinal and sacrococcygeal lesions, the average age of onset is 48 years, with a range of 8-83 years. Skull base chordomas have a mean age of onset at 44 years. There is an almost equal distribution between males and females for skull base tumours, whereas with sacrococcygeal lesions, males pre-dominate in a ratio of almost 2:1[2].

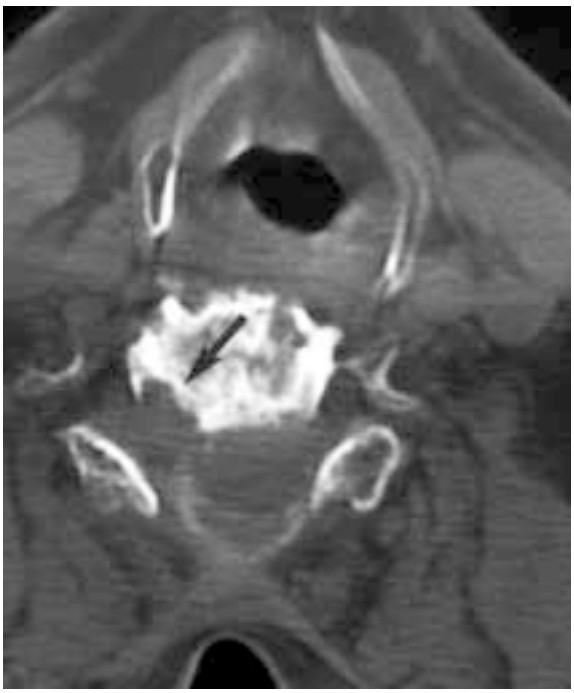

Fig. 4 Axial CT scan taken through the C5 level following a myelogram in a 73-year-old Caucasian man. Note the posterior displacement and compression of the cervical cord by a recurrent chordoma arising from the C5 vertebral body. The recurrent tumour is of soft tissue density and has extended into the right paravertebral space through an eroded right exit foramen (arrow). A midline scar indicates previous surgery with incomplete tumour resection.

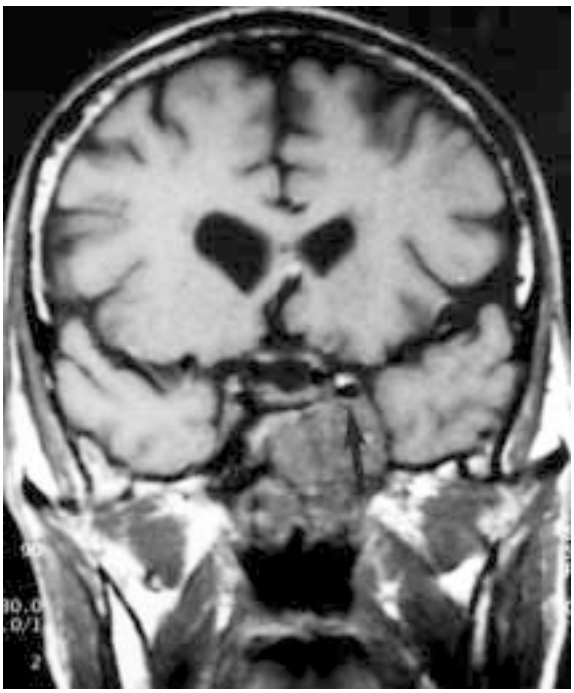

Fig. 5 Coronal T1-weighted MR image of a 68-year-old Caucasian man who presented with visual disturbance and intermittent airway obstruction for 16 weeks. The isointense mass that has partially destroyed the clivus and extended into the left cavernous sinus to partly encase the cavernous internal carotid artery (arrow). The tumour has also encroached on the nasopharynx. At surgery, 75% of the chordoma was removed.

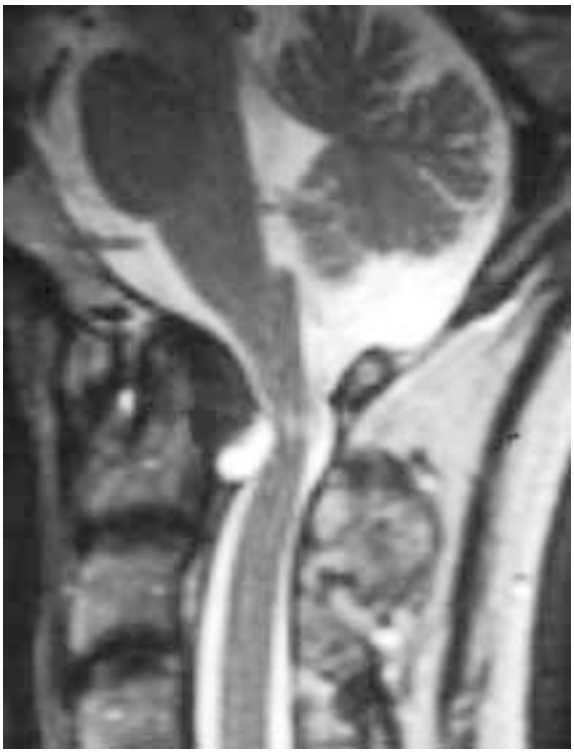

Fig. 6 Midline sagittal T2-weighted MR image taken through the craniovertebral junction of a 60-year-old Caucasian man whose main complaint was a six-week history of progressive quadriparesis. A CT scan (not shown) had shown an eroded dens. MR imaging confirms the erosion of the dens but a focal mass of which the lower portion is hyperintense is seen compressing on the cervical cord (arrow). Hyperintense signal within the cord indicates oedema/ myelomalacia. A chordoma was removed through a transoral route.

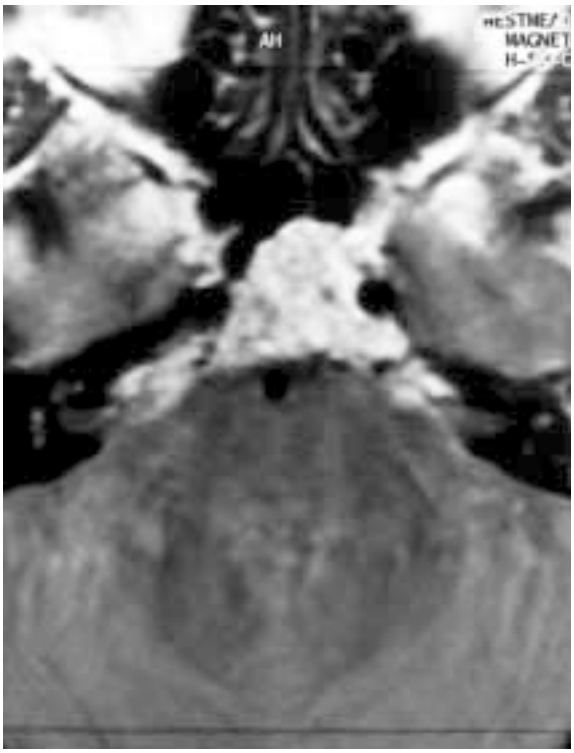

Fig. 7 Enhanced T1-weighted MR image of a 65-year-old Caucasian woman who presented with one-year history of visual impairment. The whole clivus is involved by strongly-enhancing tumour tissues in which fine internal septations are evident. There is tumour extension into the left cavernous sinus and the left internal carotid artery is partly encased. The clival component of the chordoma was excised, while the parasellar extension was treated with irradiation. The patient was well at last follow-up six years post-surgery.

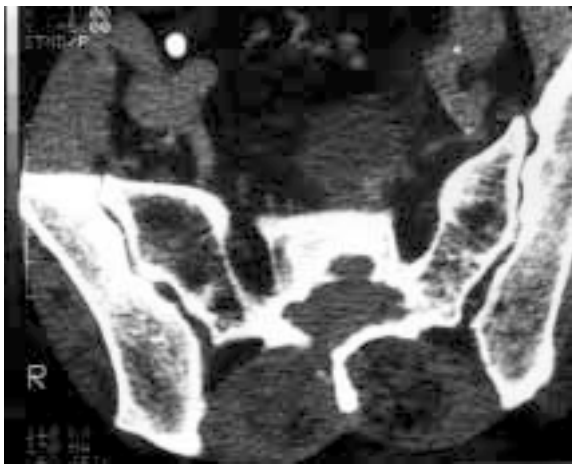

Fig. 8a Axial CT scan taken through the second sacral segment of a 64-year-old Caucasian man presenting with a 12 month history of extremity weakness and bilateral leg pains. A soft tissue tumour has expanded and eroded the sacral canal and breached the right neural arch. A tiny focus of calcification is seen. Biopsy of the tumour showed a well differentiated chordoma.

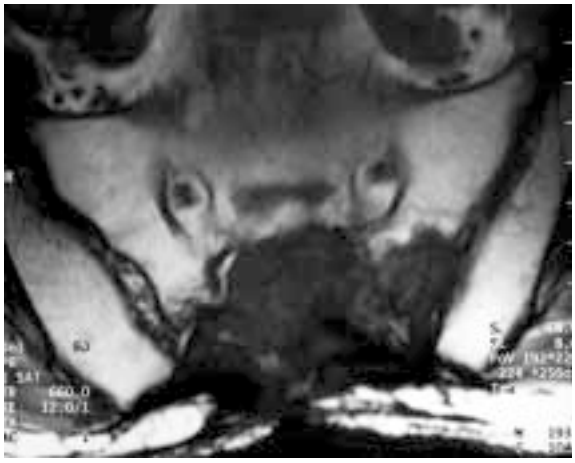

Fig. 8b Unenhanced axial T1-weighted MR image of the same patient as in Fig. 8a done three years following an incomplete resection of the tumour. There is marked tumour recurrence with signal intensity similar to the surrounding musculature.

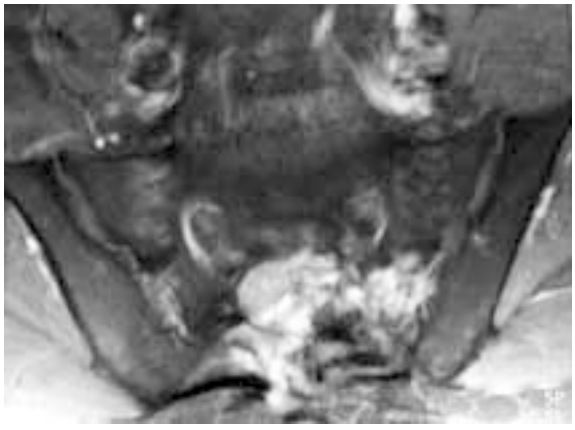

Fig. 8c Enhanced axial T1-weighted MR image taken through the same level as Fig. 8b. Note the dense but irregular enhancement of the recurrent tumour. There is distortion of the subcutaneous tissues along the midline due to scar tissue formation from previous surgery. The patient underwent a second surgical resection of the chordoma but succumbed to widespread metastatic disease a year later.

Pain in the lower back or sacrococcygeal region that is unrelieved by lying down is stated to be the dominant clinical presentation, occurring in 70% of patients in one series[3]. Other neurological complaints include leg numbness, radicular pain, and bowel or bladder incontinence[1,3]. With cranial chordomas, visual impairment due to 6th nerve palsy, headaches and facial pain from dural stretching, and lower cranial nerve irritation are the prime presentations[4].

Morphologically, chordomas are lobulated semi-translucent tumours that usually have a soft to firm consistency. They may possess a pseudocapsule and have a greyish pink appearance[4]. Multiple internal septations are found, particularly in large tumours (Fig. 2). On initial presentation, the lesional size ranges from 3 cm to 20 cm[5]. Sacrococcygeal chordomas, as in our case, are characteristically large on first presentation and may contain a considerable amount of mucin and mucoid-myxoid material within the tumour matrix. These contents explain the occasional high signal intensity shown on T1 weighted images[5]. The main conditions to be considered in the differentiated diagnoses include a solitary secondary deposit, giant cell bone tumour and neurogenic tumour (Fig. 3).

Microscopically, most cases have an admixture of epithelioid and physaliferous cells[4,6]. The latter are vacuolated cells containing intracytoplasmic mucous droplets. The physaliferous cells may be arranged in sheets, nests or lobules, and their presence characterises the tumour[6]. Importantly, foci of haemorrhage and calcifications may be present. This explains the frequently-encountered inhomogeneous appearance seen on T2-weighted MR images[7,8]. Also of importance is the presence of microscopic tumour necrosis as this is stated to be an adverse prognostic factor[6].

The typical CT imaging appearance of a sacrococcygeal chordoma is a partially-calcified moderately well-defined soft tissue tumour arising from the midline at the sacrococcygeal junction. Up to 50% of chordomas shows partial calcification[1]. Intravenous contrast enhancement on both CT and MR imaging varies from faint to dense. The excellent soft tissue resolution of MR imaging enables the inner texture of the lesion to be studied. The presence of internal septations is stated to favour a chordoma[9,10]. CT is invaluable in showing the extent of calcification and bone erosion (Fig. 4). This is particularly applicable in clival chordomas in which parasellar, intrasellar and petrous apex extensions are common.

Coronal and sagittal T1-weighted MR images are especially useful in revealing the full extent of these lesions (Figs. 5 and 6). Up to 33% of clival chordomas may present as a nasopharyngeal tumour[4]. Enhanced

T1-weighted MR scans are also useful in demonstrating tumour encasement of the internal carotid arteries when disease has spread to the cavernous sinus (Fig. 7). Whereas some of the larger clival tumours are occasionally vascular, virtually all sacrococcygeal chordomas are avascular and show no tumour circulation at pelvic arteriography[6]. In the past, pre-operative angiography was used as a road map prior to surgery but this has now been replaced by enhanced CT and MR imaging. Imaging strategy is designed to show the whole extent of lesion, enabling the selection of the best surgical approach[6,11].

Complete surgical excision is the goal of achieving a cure for tumours of the sacrococcygeal region. Preserving the S2 roots is absolutely essential for patients to retain bladder and rectal control post-surgery[12,13]. The use of CT and MR imaging has contributed to the surgical planning and successful removal of the tumour, as exemplified in this case study. Unfortunately, because of the indolent nature of the tumour with its almost silent way of bone erosion and infiltration, most lesions are not considered completely resectable by the time a firm diagnosis of chordoma is established. Current opinion favours aggressive surgery with a wide excisional margin to prevent recurrence[6,12]. This approach is adopted because incomplete removal leads invariably to recurrence, and the patients' neurological dysfunction and pain can be severe (Fig. 8). The incidence of distant metastasis increases with the presence of recurrent disease, and is estimated to be between 28%-40% in different series[6,14].

The common sites of metastases include bone, liver, lung, muscles, subcutaneous tissues and peritoneum[14]. Most investigators conclude that the use of adjuvant radiotherapy following both radical and subtotal resections improves the disease-free survival interval[15,16]. A mean disease-free survival period of 6.6 years was achieved using this regime[15]. In some instances, the biological behaviour of chordomas appears to determine survival. Watkins et al[17] analysed the survival data of 38 patients with skull base chordomas post-surgery. Two subgroups with distinct survival patterns were observed: one group with a high mortality within the first five years, and another group with an indolent disease process with near-normal life expectancy. Detailed review of the literature has identified some prognostic factors for survival. Large tumour size on initial presentation, older subjects, female patients and microscopic tumour necrosis are some of the adverse clinical indicators identified, particularly in clival lesions[6,18]. In general, there is little correlation between the morphology of the tumour and the clinical course[6]. A rare malignant variant had been described in which a sarcomatous component is present within a classical chordoma[19]. Such a tumour has a marked propensity to metastasise with a rapidly downhill course despite surgery.

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ABSTRACT

A 41-year-old man with a sacrococcygeal chordoma is presented with emphasis on its morphological and imaging features. Examples of chordomas at other sites situated along the skull base and spinal axis are illustrated. Bone erosion and expanding soft tissue mass are invariably present on both enhanced computed tomography and magnetic resonance imaging. Sacrococcygeal chordomas are usually large on initial presentation and treatment is targeted at complete surgical excision since

incomplete resection invariably leads to recurrence and distant metastases. Skull base chordomas are smaller but are less accessible to complete removal. Adjuvant radiotherapy is offered under these circumstances with the view to delay recurrence. The biological behaviour and prognostic factors for survival are summarised.

Keywords: Chordomas, computed tomography, magnetic resonance imaging, Surgery, sacrococcygeal tumour

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