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CASE REPORT

Chordoma of a thoracic vertebra
A case report

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A 51-year-old woman presented with aspecific clinical symptoms of the thoracic spine.
Radiological survey showed a tumour mass at T3-T4. Initial microscopic evaluation was suggestive of renal cell carcinoma metastasis. Lack of a primary tumour and revision of the specimens changed the diagnosis into chordoma.

Keywords : chordoma ; thoracic vertebra.

INTRODUCTION

Chordoma is a rare slow-growing malignant tumour, arising from the remnants of the chorda dorsalis (embryonic notochord). It is a locally invasive tumour and accounts for 0.2% of all CNS tumours and for 1-4% of all malignant primary bone tumours (*2,5,7,8,10,13,15,16*).

In 1857 Virchow described this tumour as *ecchondrosis physaliphora*, which in his opinion had a cartilaginous origin. This view was generally accepted until Ribbert found the same tumour during experiments in rabbits in 1895. He established its notochordal origin, so the tumours were in fact "ecchordoses", and he finally applied the term "chordoma" to this tumour (*7,10,17*).

Jallo *et al* proposed a surgical classification for vertebral spine chordomas as follows : Type I : osseous extradural lesion (majority) ; type II : extraosseous extradural lesion ; type III : osseous intradural lesion ; and type IV : extraosseous intradural lesion (*8*).

In the sacrococcygeal, sphenooccipital and vertebral region a conventional chordoma is rarely difficult to diagnose ; however, lesions outside the axial skeleton are described to yield diagnostic difficulties. Chordomas occur at any age, but they are primarily associated with middle age to late adult life. The male-female ratio is 2:1 and the vertebral chordoma mostly occurs at a somewhat younger age (*16*). About 50% of the chordomas appear in the sacrococcygeal region, 35% in the sphenooccipital region and 15% in the vertebral region (*8,13,16*). The majority of vertebral chordomas are located in the cervical spine, only 2-5% involve the thoracic

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region. Chordoma has a poor long-term prognosis, despite its slow-growing aspect and its low tendency to metastasize. The major problem is its unspecific clinical presentation. When the tumour is localized near a nerve root, symptoms of nerve root compression may arise (*14*). This is the main symptom for thoracic chordomas. We report the case of a 51-year-old woman with a thoracic chordoma. She agreed upon publication of the data concerning this case.

CASE REPORT

A 51-year-old woman presented with complaints of paraesthesia in the face and the upper thoracic area, dizziness and transpiration and other mild signs of autonomic system disorders. Furthermore she complained of vague pain in the thoracic spine region and in both legs since one year. She had an unremarkable medical history.

Examination

General and orthopaedic physical examination did not reveal abnormalities. Besides mild weakness in both legs and varying loss of sensation in the upper thoracic area there were no obvious signs of a spinal cord dysfunction.

Radiological findings

CT and MRI showed a confluent tumour mass at T3-T4 extending to the left pleural space, with mild compression of the spinal cord and involvement of the left foramen at this level (figs 1-4). A general analysis by CT and MRI scanning revealed no tumour activity elsewhere in the body.

Pathological findings

Histologic examination of a CT-guided 20G biopsy of the tumour showed morphological atypical cells. Immunohistochemical analysis showed expression on EMA, KL-1, 34BE12 and Vimentin, but no expression on CK7, CK20, Ber-EP4, CD45, Actin, Desmin, TTF-1, CA-125, S-100, oestrogen receptor and progesterone-receptor. This specific analysis most likely compared with a renal cell carcinoma metastasis, although no abnormalities as such were found at sonography and CT-scans of the abdomen.

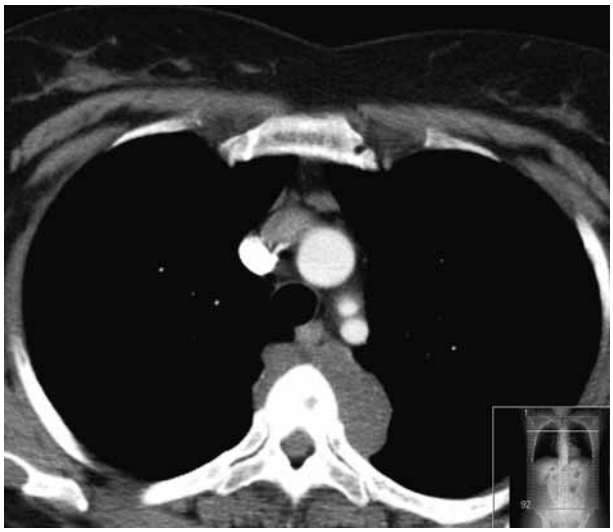

Fig. 1. — CT scan showing a tumour mass extending to the left pleural cavity.

Operation

With this diagnosis it was decided to perform a palliative anterior resection with decompression of the spinal cord. Preoperatively embolisation was used for occlusion of the nutrient arterial vessels, to prevent severe bleeding as may occur during surgery of renal metastases. Next, en bloc spondylectomy of T3 and T4 took place by an anterior approach with resection of the left second rib, followed by anterior fusion.

A Synthes®-cage (Synthes, Solothurn, Switzerland) was placed between T2 and T5 and fixation was achieved using a Z-Plate (Sofamor Danek, Memphis U.S.A.)

The cage was filled with bone from the resected second rib.

Postoperative course

The surgical procedure was complicated by a left pneumothorax which was drained successfully.

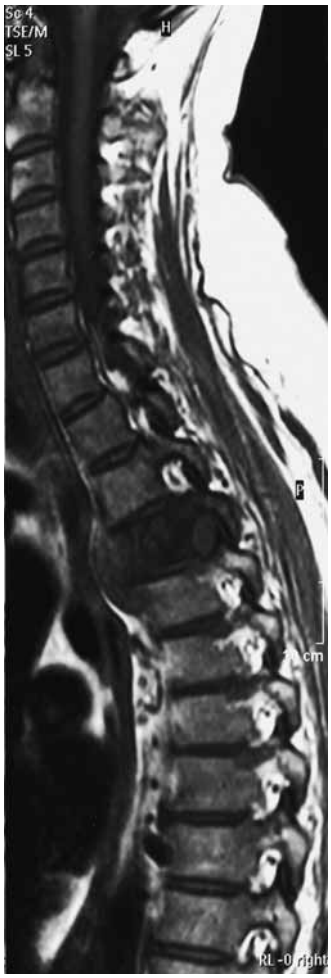

Fig. 2. — T1-weighted sagittal MRI scan showing hypointense signal in a tumour at T3 extending to the pleural cavity and involvement of the neuroforamen.

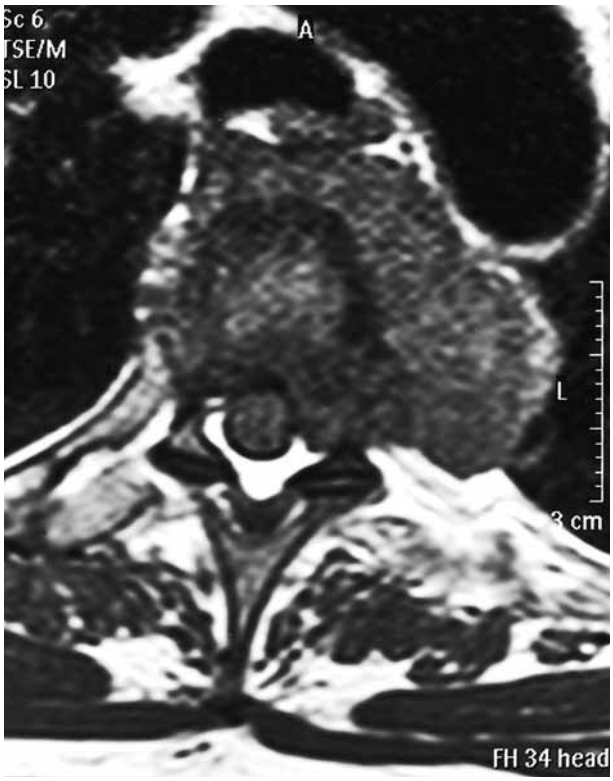

Fig. 3. — T2-weighted transversal MRI scan showing a tumour extending to the left pleural cavity with mild spinal cord compression.

Further recovery was uneventful. At two years of follow-up, the patient did not have specific complaints and she mobilised quite well, with minor neurological symptoms and varying residual pain at the site of operation. An MRI scan did not show any sign of recurrent tumour growth (fig 5).

Postoperative pathological findings

The pathologic survey of the resected vertebra showed morphological atypical cells with, in almost all cells, abundant eosinophilic cytoplasm and collagen fibers with lymphocytes influx, imposing as a renal cell carcinoma metastasis. Immunohistochemical analysis showed expression of Vimentin, EMA, KL-1, MNF116 and CAM5.2. There was no expression of S-100, hepatocyte, CEA, Actin, Desmin, CD30, CD10, CK7 and CK20 antibodies. These findings confirmed the initial diagnosis of a renal cell carcinoma.
Because of the lack of a primary renal tumour the microscopic specimens were revised once more, and ultimately the definite diagnosis was changed into vertebral chordoma.

DISCUSSION

In small biopsy samples a renal cell carcinoma closely resembles chordoma at microscopy (*12*). Coffin *et al* compared immunohistochemical features between renal cell carcinoma and chordoma biopsies. Chordomas always express on Vimentin

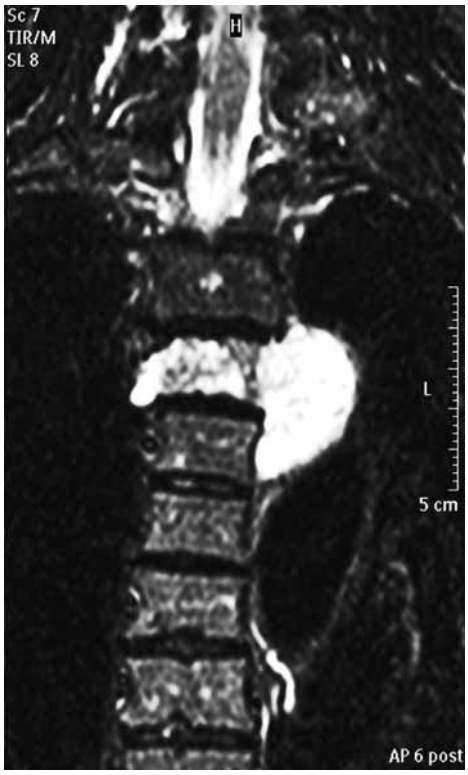

Fig. 4. — T1-weighted contrast enhanced coronal MRI scan showing a tumour at T3 extending to the pleural cavity.

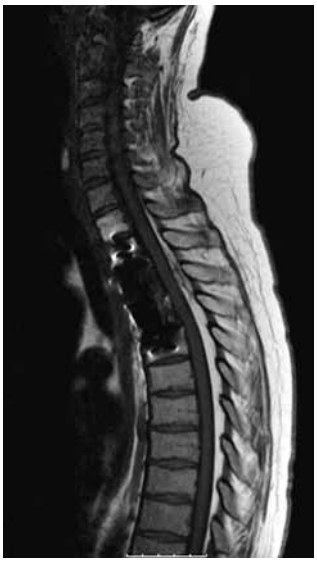

Fig. 5. — T1-weighted contrast enhanced sagittal MRI scan showing titanium artifacts, but no sign of tumour recurrence.

and mostly express on S-100 antibodies, while only 25% of renal cell carcinomas express on Vimentin and almost none reacts on S-100 antibodies (*2*). The biopsy in our patient had a positive expression on Vimentin and no expression on S-100 antibodies.

Some common and specific radiological features of chordomas of the mobile spine have been documented in other reports.

Kaulbach *et al* described the characteristics of a malignant chordoma as a consecutively expanding tumour mass originating from the chorda dorsalis, often accompanied by displacement of other organs. Osteolysis of the affected vertebra, peripheral sclerotic changes and calcification of the tumour can be recognized (*6,8*).

CT imaging is considered as the gold standard to evaluate the localization and extent of a chordoma (*3*). MRI can be helpful to identify tumour margins. The most remarkable findings of chordomas on MRI images are a high signal intensity of the tumour on T2 weighted images and enhancement of the tumour on contrast-enhanced T1 weighted images (*7*).

Treatment in general consists of surgical excision (*2*). In the mobile spine it is difficult to radically resect a chordoma, because of extradural and paraspinal extension at the time of diagnosis. When a chordoma occurs in the thoracic spine, there are two kinds of surgical treatment : piecemeal resection and en bloc resection. En bloc spondylectomy through wide bony and soft tissue margins has shown to be successful in local control of chordomas, in particular in the sacrococcygeal region. As a consequence, high sacral amputation yields the best chance of radical excision (*1,2*).

In case of a thoracic localization it is currently impossible to resect the chordoma with wide margins, as was the case in our patient. We performed en bloc marginal resection.

Local recurrence of a chordoma is more or less inevitable if excision has been marginal or intralesional. Furthermore, the recurrence rate of chordomas within two years after surgery is high in patients operated through a posterior approach, because of limited resection. Surgery through an anterior approach yields favorable results (*5*).

In the sacrococcygeal region wide margin en bloc resection has a recurrence rate of 28%, which may increase to 68% when margins are violated (*2*). Approximately 10% of the patients may expect a complete cure. Five-year survival is reported to be 78 to 84% and the ten-year survival rate reported varies from 36 to 64% (*7*).

The role of primary radiotherapy has been debated ; adjuvant radiotherapy seems to improve the local control and disease-free period after marginal resection of the tumour as in our patient (1). We started additional radiotherapy after excision of the tumour and at present, two years postoperatively, there has been no sign of tumour activity in the thoracic spine.

The incidence of chordoma metastasis in literature ranges between 3% and 48% (*1,3,5,8*). Metastatic lesions occur more often in the mobile spine than in the sacrococcygeal region. More common sites of metastasis are skin and other soft tissues, lymph nodes, lung, bone, liver and other intra-abdominal organs (*3,4,9*). Up to now, metastatic activity did not occur in our patient.

The patient in our case had an uncommon tumour of the thoracic spine, with aspecific clinical symptoms. Confusingly, the primary microscopic evaluation revealed the characteristics of a renal cell carcinoma. The definite diagnosis of an osseous extradural chordoma (type I) was not made until a secondary survey of the microscopic specimens was performed because of lack of a primary renal tumour.

In patients with spinal complaints and the radiographic appearance of a lytic lesion with a paravertebral mass of the mobile spine, chordoma should be included in the differential diagnosis.

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