Recurrent chordoma of the L2 vertebra : A case report

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Chordoma is a rare slow-growing, locally invasive primary malignant bone tumour arising from notochord remnants. It is characterised by a high local recurrence rate. Most chordomas (60%) are found in the sacrococcygeal region : only 15% originate in the mobile spine. CT-scan and MRI can help evaluate the tumour extension. En bloc resection of the tumour mass is the standard treatment. The main prognostic factor is tumour-negative surgical margins. Due to the location of spinal chordomas, adjacent to vital neural and vascular structures, this can be difficult to achieve. Therefore radiotherapy has been used when adequate excision is not possible and in case of local recurrence. We report the case of a 72-year-old female with a recurrent chordoma of the L2 vertebra, previously treated with resection and radiotherapy. The recurrent chordoma was resected using a right-sided thoraco-phreno-laparotomy as the tumour could not be resected using the left anterior approach or posterior approach due to extensive fibrosis following surgery and radiotherapy.

Keywords : chordoma ; lumbar spine ; recurrence ; case report.

INTRODUCTION

Chordomas are rare low-grade, locally invasive primary malignant bone tumours arising from notochord remnants. They have a high local recurrence rate. Most chordomas (60%) occur in the sacrococcygeal region and only 15% occur in the mobile spine (*1-3,7*). Vertebral chordomas may cause back pain with radiation to the hip, thigh, groin, knee or sacroiliacal region (*8*). We report the case of a recurrent chordoma of the L2 vertebra in a 72-year-old female.

CASE REPORT

A 72-year-old female presented with intense back pain with radiation to the right upper leg and right gluteal region. The pain intensified with movement. No neurological deficits were found on clinical examination. Sensibility, strength and reflexes in the lower extremities were symmetrical and within normal range. The patient was able to walk on her heels and toes. The straight leg raising test was negative bilaterally. CT-scan revealed an osteolytic lesion of the L2 vertebra (fig 1) extending into the peridural space on MRI-scan. It was considered to be most likely a metastasis from an unknown primary tumour. Carcinoembryonic antigen (CEA) was within normal limits. A PET-

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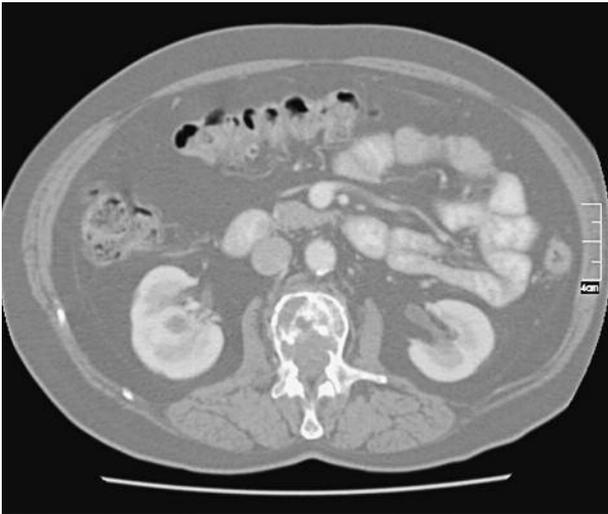 E-mail : heylensteven@yahoo.com

Fig. 1. — Pre-operative abdominal CT scan : osteolytic lesion of the L2 vertebral body with invasion of the spinal canal.

scan demonstrated an increased uptake of fluorodeoxyglucose (FDG) in the L2 vertebra and spinous process of the L3 vertebra, but showed no clues for the primary tumour and further investigations could not pinpoint a primary tumour. An open excisional biopsy was performed with a posterior decompression at the L2 level and immediate stabilization of the spine with posterior pedicle screw fixation at the L1-L3 level and posterolateral fusion with allograft bone. Histological examination of the biopsy specimen revealed fragments of haemorrhage, myxoid tissue and physaliphoric cells with immunohistochemistry positive for S100, EMA and PanCK. The cells had an eosinophilic vacuolated cytoplasm with a round to oval nucleus. Fragments of remodeling bone could also be found. The histological diagnosis was chordoma. An additional operation using a left-sided lumbotomy was performed with partial resection of the vertebral body of L2 and L1-L3 stabilization using a cage and lateral fixation. The tumour fragments, which had invaded the spinal canal, were removed. Postoperative radiotherapy with a total dose of 70 Gy was given. The patient did not develop any muscle weakness or sensibility disorder. MRI performed 2.5 years later for recurrent pain in the right upper leg showed recurrence of the disease in the posterior elements of the L2 vertebra with epidural

extension and dural compression (fig 2 & 3). The tumour reached the upper border of the right L2-L3 foramen. The tumour could not be resected using a posterior approach due to a high risk of nerve damage owing to extensive fibrosis after surgery and radiotherapy Additional radiotherapy was not an option due to the maximal doses given during the first radiotherapy treatment period. A right-sided thoraco-phreno-laparatomy was performed. A large tumour mass was found at the L2 level, parts of which had invaded the peridural space. After resection of the L1-L2 and L2-L3 intervertebral disks, the tumour mass was resected. The remnants of the vertebral body of L2 were resected, aiming for complete removal of the tumour. Due to peridural fibrosis, removal of the fragments of the tumour which had invaded the spinal canal was complex. A rib was used as a strut graft. No additional hardware was used. Histological examination of the removed tissue revealed fragments of bone and intervertebral disk tissue. Physaliphoric cells with vacuolated cytoplasm were found imbedded in a myxoid matrix. Immunohistochemistry showed the cells were positive for panCK, vimentin, EMA, but not for S100. Histological diagnosis was recurrent chordoma. Follow-up since last surgery is 10 months. The clinical and radiological evolution is satisfactory so far. Further follow-up with respect to the recurrence of the chordoma is mandatory.

DISCUSSION

Chordoma is a rare slow-growing, locally invasive primary malignant bone tumour arising from notochord remnants. The incidence is 0.5 per million (*3*). It represents only 1-4% of primary bone tumours and 6% of all malignant bone tumours (*1,6*). It is characterised by a high local recurrence rate. Chordomas can occur anywhere along the spinal column with 60% of the lesions involving the sacrum, 25% the base of the skull and approximately 15% the mobile spine (cervical, thoracic or lumbar spine) (*3,6*). In the mobile spine, cervical vertebrae are most commonly involved, followed by lumbar and thoracic vertebrae. In the cervical spine, C2 is a frequent location (*3,6*). Within the sacrum more than half of the primary bone

Figs. 2 & 3. — MRI of the lumbar spine taken 2.5 years after the initial operation, showing recurrence of the disease in the posterior elements of the L2 vertebra with epidural extension and dural compression. Imaging artefacts in the L1 and L3 vertebrae due to screws from posterior and lateral fusion of L1-L3.

tumours are chordomas. The male/female ratio is 2:1 and most chordomas are seen in late middle-age and older patients. The mean age at diagnosis is 60 years, although chordomas located at the skull base are more frequent in younger patients (*3,6*). In the lumbar spine L4 and L3 are a frequent location in the fifth decade of life in men and L3 in the seventh decade of life (*1,3*).

Vertebral chordomas may cause back pain with radiation to hip, thigh, groin, knee or sacroiliac region, especially when involving the L2 vertebra (*8*). Sensory loss, radicular pain or other neurological symptoms are more frequent in vertebral than in sacral chordomas, but these symptoms often occur only late in the evolution of the disease (*1,8*).

On plain radiographs, mobile spine chordomas may appear as lytic lesions with areas of calcification and adjacent soft tissue mass, although osteosclerosis has been described (*1,8*). CT-scan and MRI can help demonstrate the intraspinal tumoral extension (*8*). Chordomas may cause dural compression, meningeal and neural foramen invasion and vertebral destruction leading to vertebral collapse (*3,8*). Vertebral involvement is best evaluated by computed tomography, while the extent of intraspinal involvement is best assessed with MRI, as is necessary in the preoperative assessment (*1,9*). Needle biopsy sampling can be helpful in the diagnosis, but has to be carefully planned (*8*). In 25% of the cases needle biopsy of the spine fails to provide the correct diagnosis (*9*). Because chordoma is a rare tumour and the lumbar vertebra an uncommon site, it is often diagnosed on MRI or CT as a metastasis from a primary tumour that could not be found, and an open biopsy is performed. Differential diagnoses include metastasis, giant-cell tumour, chondrosarcoma and tumour-like conditions such as spinal Paget disease (*8*). Histologically chordomas are characterised by a lobular structure of physaliphoric cells with vacuolated cytoplasm, which grow trabecularly, and dual epithelial-mesenchymal differentiation (*3,6,8*). Apart from the typical chordoma as described above, there is also a more aggressive dedifferentiated high-grade variant. The chondroid variant has a better prognosis than the other chordomas (*5*).

Chordomas are considered low-grade IA or IB tumours according to the Musculoskeletal Tumour Society Staging System (*4,6*). The prognostic value of staging is therefore reduced in contrast to the importance of staging in other bone tumours. Research for other prognostic factors is therefore important, such as the location and tumour size (*4*). Chordomas of the mobile spine are locally more aggressive and more likely to metastasize than

sacral chordomas (6). The 5-year and 10-year survival rates for all chordomas are 70% and 40% respectively. Sacral chordomas have an 86% 5-year survival and a 50% 10-year survival and mobile spine chordomas have a 50% survival rate at 5 years and a 28% survival rate at 10 years (1). Chordomas are reported to metastasize late in their course to the lungs, lymph nodes, liver, bone, brain and soft tissues. Pulmonary and lymph node metastases appear most frequently followed by liver and bone metastases. The metastatic potential of chordomas may have been underestimated in the past (2,5,7). Metastases have been reported in 80% of vertebral chordomas and 43% of all chordomas, but reported frequencies vary considerably in the literature (5,7). Survival is related more to local tumour progression than to metastasis (2,5,6). Early radical surgical excision of the tumour is therefore the only curative treatment (3,6-8). Due to adjacent vital neural and vascular structures this is often difficult, nonetheless a more aggressive surgical approach has evolved from intralesional partial excision to "en bloc" resection. A disease-free margin can be achieved in only half of the sacrococcygeal chordomas treated with surgery. This figure is even less for chordomas of the mobile spine or skull-base. For chordomas of the mobile spine involving both anterior and posterior vertebral components a two-stage anterior and posterior approach has been suggested including tumour resection, immediate stabilisation and spine reconstruction using replacements or bone grafts with internal fixation (1,6,7,9). Radiotherapy may provide short-term benefit when adequate excision is not possible and in case of local recurrence (1,3). Despite improved treatment, local recurrence rates remain high (7). The most important prognostic factor for local failure is the surgical margin (5). Other prognostic factors may be tumour necrosis, tumour size and the performance of invasive diagnostic procedures outside of a tumour center (9). Chordomas are considered to be radioresistant with a curative dose of 70 Gy or more. Because of the location of chordomas, the tolerance dose of surrounding tissue is often less than the curative dose. However, because of the difficulty in obtaining disease-free margins, radiotherapy has

lately been researched more and more (5,9). With the development of more sophisticated technology, such as intensity modulated radiation therapy (IMRT), proton beam radiation therapy, hadron radiation therapy alone or in combination with conventional photon beam therapy and radiosurgery, the overall target coverage has been improved to doses reaching the curative dose (2,5,10). Uncertainty remains whether to apply radiotherapy only when resections have been incomplete or as a standard post-surgery treatment. While the dedifferentiated chordoma variant may be sensible to certain chemotherapeutical agents, the classical chordoma variant seems to be non-responsive to chemotherapy, although reports of tumour response do exist, but the response rates are low (2). Recently there has been an interest in the use of Imatinib, which has been described to have antitumour activity (2,5,9).

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