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Chordoma of the lumbar L2 vertebra: case report and review of the literature

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Abstract We report four new cases of chordoma of "the mobile spine", all at the L2 level. Diagnosis was often delayed due to predominantly nonspecific low back symptoms; however, neurological involvement is more frequent than in chordoma with a sacrococcygeal localization. No pathognomonic images have been described for any imaging modality, and differential diagnosis should include metastases, chondrosarcoma, and giant-cell tumor. Histopathological analysis can be performed on CT-guided puncture biopsy samples, but a high level of suspicion must be present and, if there is any doubt, immunohistochemical studies should be carried

out. Despite being the treatment of choice, complete tumor resection by a double-approach spondylectomy is barely feasible at the L2 level.

Key words Chordoma · Lumbar spine · Surgical treatment

Introduction

Chordoma is a slow-growing, locally invasive, rare tumor of the adult spine, that rises from and may develop at any point along the original notocordal track, but it is usually noted at the rostral and caudal ends (e.g., the basiocciput and the sacrococcyx) [24, 29]. However, its occurrence in the mobile spine has been well documented, with 33% of cases involving the lumbar spine [5].

We present four new cases of chordoma located in the upper lumbar spine (L2).

Case reports

Case 1

A 50-year-old white man was first examined 1 year after the onset of progressive low back pain. At admission bilateral lower limb

paresthesias and evident gait disturbance were found. Plain radiographs and CT scans showed a lytic lesion involving the vertebral

Fig. 1 T1-weighted MR image of case 1 showing L2 body bone marrow replacement with a hypointense striated pattern. Epidural and paravertebral involvement are also seen

Fig. 2A,B Case 2. **A** Lateral plain lumbar spine film showing posterior scalloping of the L2 body with pedicle erosion, which increases the neural foramen diameter. **B** Sagittal T1- (left) and T2-weighted MR images showing a large tumor mass emerging from L2 and involving completely the spinal canal, with a hypointense T1 signal and hyperintense T2 one

Fig. 3A,B Case 3. **A** Frontal plain lumbar spine film. Speckled L2 body sclerosis and irregular cortical bone is characteristic though not pathognomonic. Pedicle involvement and pathologic fracture are also evident. **B** CT-scan of guided percutaneous L2 body biopsy. Retroperitoneal tumoral spreading behind the inferior cava vein is evident

Fig. 4 Post-laminectomy MR image of case 4 showing iatrogenic kyphosis and tumoral involvement of L1 and L2

body and both pedicles of L2, with paravertebral soft tissue and epidural space extension. MRI revealed cauda equina root compression (Fig. 1). CT-guided puncture biopsy was performed. The histopathological diagnosis was chordoma. The patient refused surgical treatment and was lost to follow-up.

Case 2

A 66-year-old white woman suffered night low back pain beginning 2 years before diagnosis and progressively increasing. At admission she showed left limb numbness, crural paresis, and positive straight leg raising test. CT and MRI showed L2 body involvement with paravertebral soft tissue mass; epidural space extension at the L2 and L3 levels, and L2 pedicle destruction were evident (Fig. 2). CT-guided biopsy revealed a chordoma. Double-approach resection was planned with, firstly, a posterior approach (L2-L3 laminectomies, tumor resection, and T11-L5 Hartshill instrumentation). Unfortunately cardiac arrest occurred at the end of the posterior approach and the patient died after surgery.

Case 3

A white 57-year-old woman with severe low back pain radiating to anterior region of both thighs presented with paresthesias and numbness that had started 8 months prior to admission. Pain increased with L1-L2 spinous process percussion. Plain radiographs showed L2 body osteolysis with pathological fracture and left L2 pedicle involvement (Fig. 3A). Technetium 99m-HMDP scintigraphy revealed an L2-L3 hot spot. CT also showed a retroperitoneal soft tissue mass extension. Scanner-guided biopsy (Fig. 3B) suggested a clear-cell kidney carcinoma metastasis, but as doubt persisted, immunohistochemical studies were performed and chordoma was categorically recognized (positive S100-protein, positive keratins, negative carcinoembryonic antigen). An anterior tumor resection, methylmethacrylate vertebral reconstruction, and Malaga anterior system stabilization were performed by one of the authors (P.M.). One month later, a posterior resection, stabilization with Lea Plaza frame, and posterolateral fusion completed surgical treatment. Radiotherapy up to a total dose of 46 Gy was added. One year later the patient was asymptomatic, with an acceptable level of activities and a stable spine, and she was free from disease.

Case 4

A 70-year-old white man with chronic low back pain, which had increased 1 year before initial surgical treatment, was first operated on by another surgeon who performed a laminectomy due to a lytic L2 lesion. Histopathology revealed chordoma and postoperative radiotherapy was indicated.

Ten months later he was seen by one of the authors (N.F.), suffering severe low back pain, bilateral thigh pain, and Frankel C paraparesis. Plain radiographs showed L2-L1 involvement, wide laminectomy, and secondary kyphotic deformity. CT and MRI confirmed vertebral body destruction as well as large epidural extension (Fig. 4). Posterior partial resection combined with neural release and T9-L5 Lea Plaza instrumentation was performed. Pain was substantially reduced and the patient completely recovered from a neurological standpoint. Ten months later relapse occurred and an anterior approach with partial resection, iliac strut graft, and T9-L4 Malaga system stabilization was performed. The patient improved to Frankel D, with pharmacological pain control. One year later disease progression led to death.

Discussion

Approximately one-sixth of chordomas arise in the “mobile spine” [33], this means the cervical, thoracic, or lumbar spine, with the latter involved in 33% of cases. Clinical findings in these four new reported cases are similar to those previously reported by Bjornsson from the Mayo Clinic files [5], but the new cases all had an L2 location.

No difference was found regarding mean age and symptom duration before surgery between this series and that previously reported [5], but the male:female ratio was different (1:1 vs 2:1).

The main complaint associated with vertebral chordomas is pain, and when it involves the L2 vertebra, radiation to hip, thigh, groin, knee, or sacroiliac region is not infrequent [33], as in cases 3 and 4. Even though radicular pain, sensory loss, or other neurological deficits are present in 10% of chordoma cases, these are more common in chordomas of the mobile spine than of the sacrococcygeal region [5, 18, 33],

Lumbar chordomas are usually diagnosed late due to rare location, slow growth and nonspecific image appearance. Nevertheless, newer imaging techniques may allow earlier diagnosis [2, 9, 33].

Though upper lumbar spine involvement is certainly infrequent [5], the imaging studies show similar findings to chordomas of the sacrococcygeal and spheno-occipital regions. They appear predominantly as lytic lesions on plain radiographs, with areas of amorphous calcification and adjacent soft tissue mass [2, 25, 29]. Vertebral collapse may occur (Fig. 3A). A blastic component may be predominant [9]. Differential diagnosis includes other primary or secondary spine tumors like metastasis, giant-cell tumors, and chondrosarcomas [29, 33], and tumor-like conditions such as spinal Paget disease [16].

As it originates in notocord remnants, chordoma usually shows a radiating infiltrative pattern involving epidural space and prevertebral and lateral paravertebral regions. It can cause dural compression and eventually meningeal sheath invasion and cerebro-spinal fluid spreading (drop metastases) [25]. Extension into the neural foramen leads to its erosion, giving images similar to neurofibromatosis, as in case 2 (Fig. 2A).

During retroperitoneal spreading, psoas muscle and great vessels are displaced. Adjacent disk spaces and vertebral bodies can be involved (Fig. 4). The extent of vertebral destruction is better demonstrated on CT scans. However, MRI and myelo/CT scans are more suitable for establishing the extent of intraspinal involvement.

Owing to a high free-water content, T1-weighted MR images appear hypo- or isointense to spinal cord images. T2-weighted images appear as a hyperintense signal, usually with a striated pattern at the vertebral bodies, with pseudocapsule and strands in soft tissue extensions [2] (Fig. 2B).

Although bone technetium 99m-HMDP and gallium scintigraphy may be of some value [32], unfortunately neither single-imaging diagnosis method can confirm suspicion of chordoma, or distinguish its type (typical or chondroid) [34].

Grossly, it is a lobulated, grayish, soft mass that may have the appearance of mucous carcinoma or chondrosarcoma [18, 26, 33]. Microscopically, it shows a distinct lobular architecture formed by two cell types: physaliphorous cells with ample vacuolated cytoplasm (mucin-containing vacuoles) and "signet ring" type cells, both separated by a variable amount of mucoid intercellular tissue [26, 29, 33].

However, histopathological analysis, particularly with samples obtained by closed puncture, is not a simple task when chordoma is not suspected because of a rare location [4]. Nevertheless, closed biopsy is probably the most suitable technique for the spine [23, 30, 31], especially when it is CT guided [3]. In those cases, immunohistochemical studies are certainly helpful in distinguishing chordoma from adenocarcinoma and cartilage tumors [1, 5, 14, 17, 19, 27–29], as in case 3.

Despite some controversial statements about chemotherapy [7, 10] and radiotherapy [4, 5, 13], there is general agreement that complete surgical "en bloc" resection with a wide margin is the treatment of choice in chordomas [17, 33, 35]. In the mobile spine, when both anterior and

posterior vertebral components are involved, a staged complete spondylectomy must be preferred, including an anterior approach for tumor resection and spine reconstruction followed by a posterior approach for completion of resection, arthrodesis, and stabilization [4, 5, 33].

Anterior methylmethacrylate reconstruction after vertebral tumor resection and posterior segmental instrumentation have already proved their usefulness in spinal neoplastic conditions [6, 8, 11, 12, 15, 20–22, 36].

Even so, at the L2 level wide resection is barely feasible [4], and it is usually intralesional. A careful resection of all gross tumor by a combined approach in these cases may achieve an almost complete resection [4, 33].

Conclusions

1. L2 chordomas are rare, with long-standing symptoms prior to diagnosis. A high suspicion rate should be present when dealing with a patient who shows pathological non-specific image studies.
2. MR images though not pathognomonic, may help reduce diagnostic delay.
3. CT-guided biopsy, with immunohistochemical sample studies, should be thoroughly performed.

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