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Case Report

Spinal Chordoma: An Unusual Radiological Presentation: A Case Report

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Abstract

We present a case of a chronic and undetermined low back pain in an elderly female patient. The MRI was crucial to characterize a high cellularity mass involving the vertebral body and the surrounding structures in a misleading diagnosis.

A discussion of the imaging findings and differential diagnosis of a rare case of spinal Chordoma in a high lumbar position (L2) is provided as follow.

Keywords: Chordoma; Magnetic resonance; X-Ray; Spine; Lumbar

Case Presentation

A 74-year old woman, with no medical history of interest, presented to our department with a 3-month history of low back pain, not responsive to pain-killing therapy. The patient, therefore, underwent lumbar radiography in the Antero Posterior and Lateral view.

The plain film displayed a heterogeneous lytic mass of the L2 vertebral body, extending into the disc space and probably involving also the pedicles on the right side. The neural foramen appeared to be not enlarged (Figure 1).

Consecutively, a MRI of the spine was performed at 1.5 T (Achieva, Philips, The Netherlands), using a 16-channel body phased array surface coil.

MRI analysis was performed with T1 and T2-weighted Turbo Spin Echo, Inversion Recovery, and Gradient Echo sequences, with axial, coronal and sagittal planes, with and without fat saturation, completed with sequences after intravenous administration of 0,1 mg pro kilo of paramagnetic contrast media (Gd-DTPA).

The presence of an expansive mass, of 5 x 6 x 4,5 cm, with irregular edges, was confirmed (Figure 2 sagittale).

This showed heterogeneous hypo to isointense signal in the T1 weighted images, hyper intense signal to cerebral spinal fluid in the long TR sequences, with some inner spots of low signal in all the impulse sequences, probably due to calcifications (Figure 3). After intravenous administration of contrast agent, a slight and uneven enhancement was observed within the lesion (Figure 4).

The mass took up the right side of the spinal canal, mostly in the back wall of L2, compressing the dural sac and partially occluding the intervertebral foramina between L2 and L3.

The bulk was involving also the omolateral vertebral pedicle, the correspondent articular process and the paravertebral soft tissue, pushing back the iliopsoas muscle.

The suspect of a primary bony tumor was supposed but wasn't excluded a possible replication from an unknown neoplasia.

A total body CT scan with and without contrast was performed to find a possible primary tumor or, eventually, to stage the pathology: no replications were found. The suspect of a primary lesion was then confirmed (Figure 5 CT).

Due to instable clinical conditions of the patient and the local infiltration of the tumor, the surgical planning was limited to a palliative approach. Vertebral stabilization of the spine was executed from D12 to L4, along with a right hemilaminectomy of L2. An

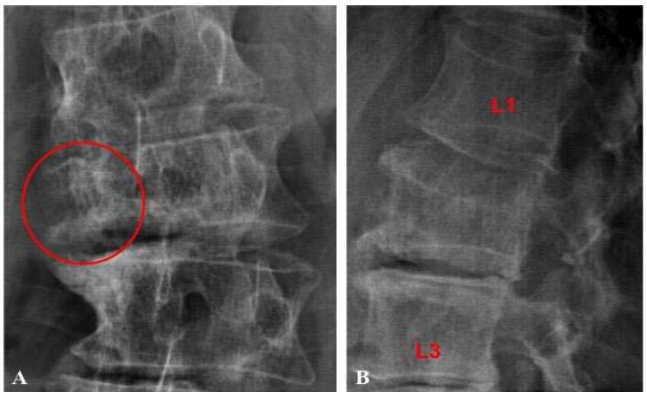

Figure 1: Antero-Posterior (A) and lateral (B) plain lumbar spine film. Mottled L2 body sclerosis and irregular cortical bone is seen on the right side. A possible pedicle involvement and pathologic fracture are also evident (circle). No enlargement of the neural foramen diameter was documented.

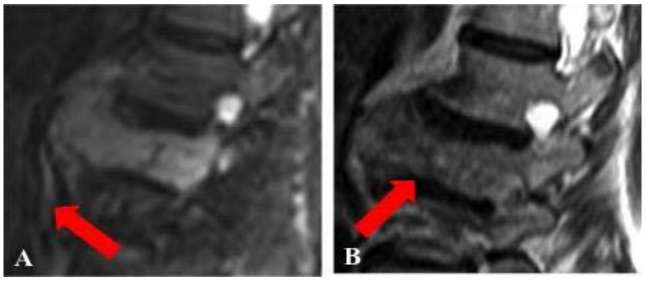

Figure 2: Sagittal STIR (A) and T2 (B) weighted images illustrate a hyper intense, ovalar mass infiltrating the L2 vertebral body. Note the extension of the tumor in the anterior and right paravertebral space (arrow in A).

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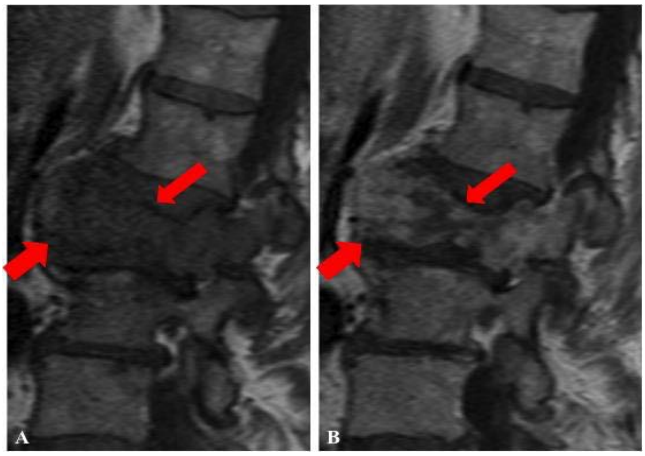

Figure 3: Sagittal pre (A) and post contrast (B) T1 weighted images. Heterogeneous hypo to isointense bulky mass in the pre contrast images, with slight enhancement after gadolinium injection. Some inner L2 body small areas of low signal, probably due to calcifications, are seen (arrows in B).

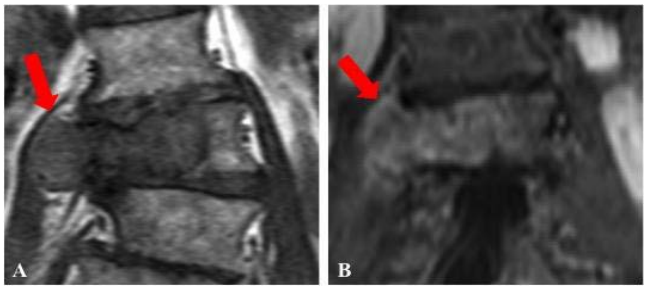

Figure 4: Coronal T1 (A) and T1 SPIR after contrast agent (B) weighted images confirming the paravertebral extension of a growing mass of the L2 body with in homogenous enhancement, pushing aside the ileopsoas muscle.

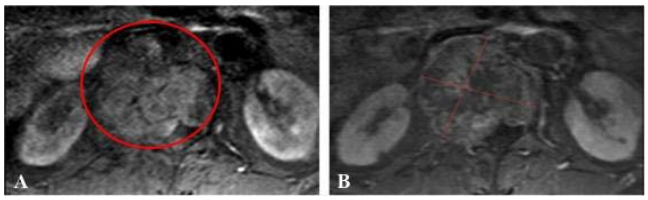

Figure 5: Axial T1 (A) and T1 SPIR after contrast agent (B) weighted images. The bulk is involving also the right vertebral pedicle, the correspondent articular process and the paravertebral soft tissue.

Intraoperative biopsy was carried out.

No secondary implants were found during the procedure.

A lumbar blank CT with bony reconstructions was performed after the surgical treatment, which displayed the persisting mass and the decompression of the neural structures (Figure 6 CT2).

Pathological examination revealed the presence of a 5,2 × 6 cm Chordoma with areas of reactive ostheogenesis within it (Figure 7&8).

During the multidisciplinary meeting, in mutual agreement with the oncologist and the radiotherapist, no further treatments were suggested due to the weak clinical condition of the patient and the location of the tumor. Unfortunately the patient refused chemotherapy.

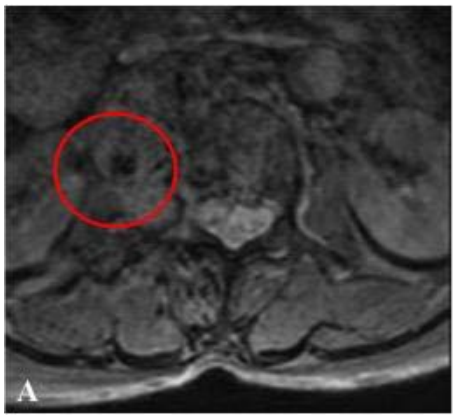

Figure 6: Axial T2 weighted image with particular of a hypo intense areola within the mass suggestive for bony fragment due to the pathologic fracture (circle).

The patient was dismissed on the tenth day post-operatively with a good clinical process and completely recovered from a neurological standpoint.

Discussion

Chordoma are uncommon aggressive extradural lesions of the bony spine (being part of the sarcoma's family), arising from remnants of the primitive notochord and representing approximately 3 to 5 percent of primary bone tumors, with an incidence of 0.08/100.000 inhabitants [1].

Despite Chordoma is considered to be a slow growing tumor it can recur very frequently, mostly because the nearness with neurovascular bundles, presenting also a malignant transformation [2-5].

They usually grow into the sacrococcygeal bones (50%) and in the basi-sphenoidal bones, especially in the clivus (30-40%), but other locations are described in literature mostly into the vertebral bodies [6-8].

It has a higher prevalence for male with a ratio of 2:1 and it is usually seen in adults (30-70 years) with a peak around 50 years for the sacrococcygeal localization [9].

Chordoma presents three different subtypes: typical ("classic"), chon droid and dedifferentiated.

Classic Chordoma consists mainly of mucin and glycogen

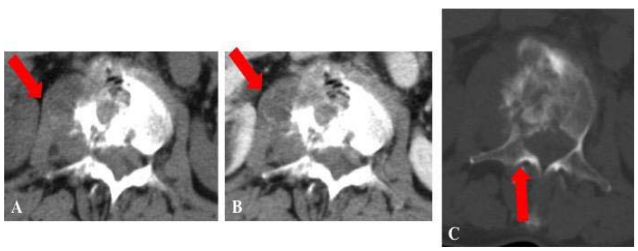

Figure 7: Axial pre (A) and post contrast (B) CT slices before the surgical treatment, showing the infiltration of the dysplastic tissue into the bone. Right paravertebral extension (arrows in A and B) with omolateral pedicle involvement (arrow in C). Note the bony osteolysis of the L2 body.

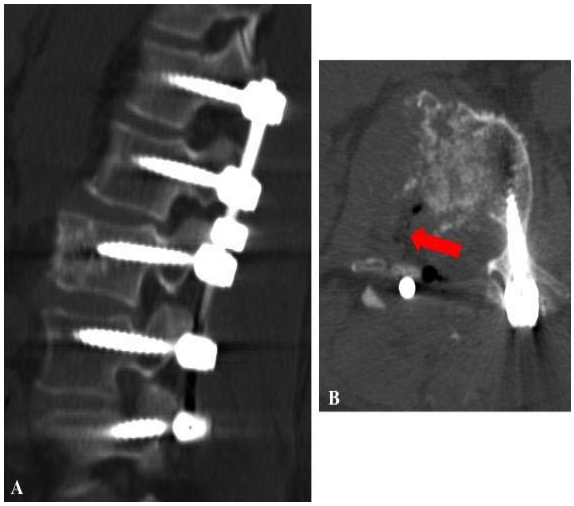

Figure 8: Blank CT with bone filter sagittal reconstruction (A) showing the vertebral stabilizer applied between D12 and L4. In the axial projection (B) result of the right laminectomy (arrow).

vacuoles. Chon droid subtype have mostly stromal elements while the undifferentiated Chordoma with sarcomatous behaviour seems to be the most aggressive, growing especially in the sacrococcygeal region [1,10-12].

The lumbar Chordoma is an uncommon location and only few cases are described in literature, even if its appearances are similar to those in the sacrococcygeal and spheno-occipital regions [5,10,13-17].

Indeed, they appear frequently as lytic lesions both on plain films and CT scan, with areas of irregular calcifications and paravertebral soft tissue. T1-weighted MR images display a hypo-isointense to spinal cord signal. T2-weighted images appear as in homogenous hyper intense signal for the high free-water content, with some hypo intense areas within it, owing the calcifications [18,19].

The unfrequent location and the a specific appearance of this tumor can confound the diagnosis, leading the radiologist to more frequent causes of lumbar pain owed by vertebral masses, such as metastases or other primary tumors.

With these limitations, we tried to point out the different imaging features to do not miss this kind of lesion; the possible differential diagnosis includes:

- Chondrosarcoma: even if it is extremely rare in the spine, on plain films it can cause lytic destruction and calcified matrix in the form of radio dense swirls, rings or arcs [18]. It can be associated with a soft tissue mass, as well. MR signal intensity of Chondrosarcoma is heterogenous: focal areas of decreased signal intensity on T2-weighted images due to prominent calcifications, or high signal intensity representing cartilage.
- Lymphoma: generally presents as a multifocal disease with sclerotic lesion ("ivory vertebra"). Moreover, it can arise from a loco regional lymphoadenopathy.
- Plasmacytoma or myeloma: normally appears with a typical lytic lesion ("salt and pepper" behaviour) owing the high cellular turn over.
- Giant cells tumor shows a geographic distribution and lytic lesions, rarely with a sclerotic border
- Aneurismatic Cyst: appears as a typical blood filled lesion, clearly displayed with the MR sequences sensitive for susceptibility magnetic artifacts
- Lythic Metastasis: despite as the Chordoma usually present hypointesnse at T1-w images with a variable hyper intense signal inT2-w sequences they often are multiple, involving bodies and posterior elements [1,11,18,19].

Focusing back to our case, we found a rare case of L2 Chordoma with misleading findings: owing the presence of areas of low intensity signal in all the MR pulse sequences, a chon droid origin of the Chordoma could be suspected. Nevertheless, the pathologist excluded it, confirming the presence of areas of reactive osteogenesis within the tumor tissue. The small hypo intense areolas, confirmed as high dense spots with the CT, could resemble calcifications within the tumor lesion or a chon droid matrix. Conversely, the histologic examination assessed those fragments as pathologic vertebral fracture.

According to Jung et al. MRI findings suggestive of metastatic compression fractures are as follow: convex posterior border of the vertebral body, abnormal signal intensity of the pedicle or posterior element, epidural mass, encasing epidural mass, focal paraspinal mass and other spinal metastasis [20]. Despite these features, we learnt that other elements need to be considered to achieve a correct diagnosis: a silent history for tumor in a 74 old female patient with chronic low back pain should let us thinking to other possible causes than metastasis. However, the MRI was the most important tool to guide the following choices: T1 weighted sequences helped us to exclude the involvement of the cord and provide a baseline for comparison with the post-gadolinium images.

The contrast agent was necessary to study the vascular behaviour of the mass and to demonstrate the viable tumor boundary. The sequences sensitive for fluid helped us to recognize the mucoid matrix of this kind of tumor which is part of the sarcoma family [21].

DWI could be use full in differentiate malignant to benign vertebral fractures but in our case there were no pathognomonic sign could help us to characterize the solid mass [22-25].

Conclusion

In conclusion we discussed a not common location of Chordoma with misleading radiologic features where the MRI helped defining the relationships between the tumor, the adjacent structures and its typical signal intensity. However the differential diagnosis between metastasis and other primary bone tumors is still a tough issue.

Our porpouse was to highlights the radiologic and MR characteristics of this rare tumor in a even more rare position in order to keep in mind the Chordoma in the diagnostic process.

References

1. TJM. Magnetic Resonance Imaging of the Spine. 2nd edition. 1994.
2. Vujovic S, Henderson S, Presneau N, Odell E, Jacques TS, Tirabosco R,

et al. Brachyury, a crucial regulator of notochordal development, is a novel biomarker for chordomas. J Pathol. 2006; 209: 157-165.

3. Launay SG, Chetaille B, Medina F, Perrot D, Nazarian S, Guiramand J, et al. Efficacy of epidermal growth factor receptor targeting in advanced chordoma: case report and literature review. BMC Cancer. 2011; 11: 423.

4. Ishida T, Dorfman HD. Chondroid chordoma versus low-grade chondrosarcoma of the base of the skull: can immunohistochemistry resolve the controversy? J Neurooncol. 1994; 18: 199-206.

5. Boriani S, Bandiera S, Biagini R, Bacchini P, Boriani L, Cappuccio M, et al. Chordoma of the mobile spine: fifty years of experience. Spine (Phila Pa 1976). 2006; 31: 493-503.

6. Royo Crespo I, Rivas de Andrés JJ, Embún Flor R, Cortés Franco S. Chondroid chordoma in an atypical location. Arch Bronconeumol. 2013; 49: 491-493.

7. Smolders D, Wang X, Drevelengas A, Vanhoenacker F, De Schepper AM. Value of MRI in the diagnosis of non-clival, non-sacral chordoma. Skeletal Radiol. 2003; 32: 343-350.

8. Williams SP, Beckerman B, Piña Fonti ME. A 69-year-old presenting with musculoskeletal low back pain: a case of lumbosacral chordoma. J Chiropr Med. 2014; 13: 144-148.

9. Farsad K, Kattapuram SV, Sacknoff R, Ono J, Nielsen GP. Sacral chordoma. Radiographics. 2009; 29: 1525-1530.

10. Manzone P, Fiore N, Forlino D, Alcala M, Cabrera CF. Chordoma of the lumbar L2 vertebra: case report and review of the literature. Eur Spine J. 1998; 7: 252-256.

11. Osborn AG. Osborn's brain: imaging, pathology, and anatomy. 1st edition. Salt Lake City, Utah: Amirsys Pub. 2013.

12. York JE, Kaczaraj A, Abi-Said D, Fuller GN, Skibber JM, Janjan NA, et al. Sacral chordoma: 40-year experience at a major cancer center. Neurosurgery. 1999; 44: 74-79.

13. Bjornsson J, Wold LE, Ebersold MJ, Laws ER. Chordoma of the mobile spine. A clinicopathologic analysis of 40 patients. Cancer. 1993; 71: 735-740.

14. Breen N, Eames N. Chordoma of the lumbar spine - a potential diagnosis not to be forgotten. J Surg Case Rep. 2012; 4.

15. Nishiguchi T, Mochizuki K, Tsujio T, Nishita T, Inoue Y. Lumbar vertebral chordoma arising from an intraosseous benign notochordal cell tumour: radiological findings and histopathological description with a good clinical outcome. Brit J Radiol. 2010; 83: 49-53.

16. Sartoretti-Schefer S, Oberle J, Binkert C, Pfofe D, Gutzeit A. Lumbar vertebral chordoma: diffusion-weighted imaging findings are helpful to narrow the differential diagnosis. A case report. Clin Imag. 2013; 37: 176-179.

17. Utne JR, Pugh DG. The roentgenologic aspects of chordoma. Am J Roentgenol Radium Ther Nucl Med. 1955; 74: 593-608.

18. Atlas SW. Magnetic resonance imaging of the brain and spine. 3rd edition. Philadelphia, Pa; London: Lippincott Williams & Wilkins. 2002.

19. Resnick DNW. Diagnosis of bone and joint disorders. Saunders ed.1988.

20. Jung HS, Jee WH, McCauley TR, Ha KY, Choi KH. Discrimination of metastatic from acute osteoporotic compression spinal fractures with MR imaging. Radiographics: a review publication of the Radiological Society of North America. 2003; 23: 179-187.

21. Romeo S, Dei Tos AP. Soft tissue tumors associated with EWSR1 translocation. Virchows Arch. 2010; 456: 219-234.

22. Baur A, Huber A, Ertl-Wagner B, Durr R, Zysk S, Arbogast S, et al. Diagnostic value of increased diffusion weighting of a steady-state free precession sequence for differentiating acute benign osteoporotic fractures from pathologic vertebral compression fractures. Am J Neuroradiol. 2001; 22: 366-372.

23. Castillo M, Arbelaez A, Smith JK, Fisher LL. Diffusion-weighted MR imaging offers no advantage over routine noncontrast MR imaging in the detection of vertebral metastases. AJNR Am J Neuroradiol. 2000; 21: 948-953.

24. Maeda M, Sakuma H, Maier SE, Takeda K. Quantitative assessment of diffusion abnormalities in benign and malignant vertebral compression fractures by line scan diffusion-weighted imaging. AJR Am J Roentgenol. 2003; 181: 1203-1209.

25. Zhou XJ, Leeds NE, McKinnon GC, Kumar AJ. Characterization of benign and metastatic vertebral compression fractures with quantitative diffusion MR imaging. AJNR Am J Neuroradiol. 2002; 23: 165-170.

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