

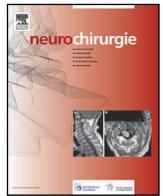


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Short clinical case

An odd and serious “disc bulging”!

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ABSTRACT

Background and importance. – Cauda equina syndrome is a common acute medical condition, usually caused by large degenerative disc herniation or metastatic lumbar disease. We describe a patient who presented with a lesion featuring both discal and tumoral characteristics.

Clinical presentation. – A 41-year-old woman presented with ongoing back pain, progressive lower-limb weakness and sphincter disorder. Magnetic resonance imaging showed a very odd-looking large anterior epidural lesion originating from the L3–L4 space and severely compressing the roots of the cauda equina. Partial surgical decompression was performed in emergency. At a later time, redo surgery was performed to maximize resection, and was unfortunately followed by several complications. After 6 surgical procedures including a ventriculo-peritoneal shunt insertion and intensive rehabilitation, the patient could walk independently with the aid of one crutch. Following collegial review, the diagnosis of low-grade chondrosarcoma of the intervertebral disc was suggested.

Conclusion. – We report on a very unusual and therapeutically challenging spinal tumor diagnosed as low-grade chondrosarcoma of discal origin, an entity never previously described.

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1. Abbreviations

CSF	Cerebrospinal fluid
CT	Computed tomography
MRI	Magnetic resonance imaging
WHO	World Health Organization

2. Background and importance

Cauda equina syndrome is a common acute medical condition, usually caused by large degenerative disc herniation or metastatic lumbar disease. Here we describe a patient who presented with paraparesis due to a lesion featuring both discal and tumoral characteristics. We discuss treatment and results in the light of the literature.

3. Clinical presentation

3.1. Initial presentation

A 41-year-old woman living in the Republic of Cabo Verde, with no significant medical history, was suffering from progressive cauda equina syndrome. In Cabo Verde, she underwent lumbar computed tomography (CT), which showed no obvious lesion. In time she could barely walk and, due to the limited medical resources available locally, she came to France.

A redo CT scan was performed (Fig. 1) before transfer to our institution. Magnetic resonance imaging (MRI) showed a large anterior epidural lesion originating from the L3–L4 disc, severely squeezing the cauda equina (Fig. 2). It was decided to perform surgical decompression urgently.

3.2. First surgical procedure, results and progression

After a standard laminectomy, a tumor of mixed soft and friable texture, quite easily resectable and moderately bloody, was progressively removed (see video). In some areas, it seemed to invade the dura mater, which was thinned. Hermetic closure was achieved with fat superimposition. No cerebrospinal fluid (CSF) leakage occurred and after three days' bed rest, she was able to rise. Postoperative MRI showed significant residual tumor.

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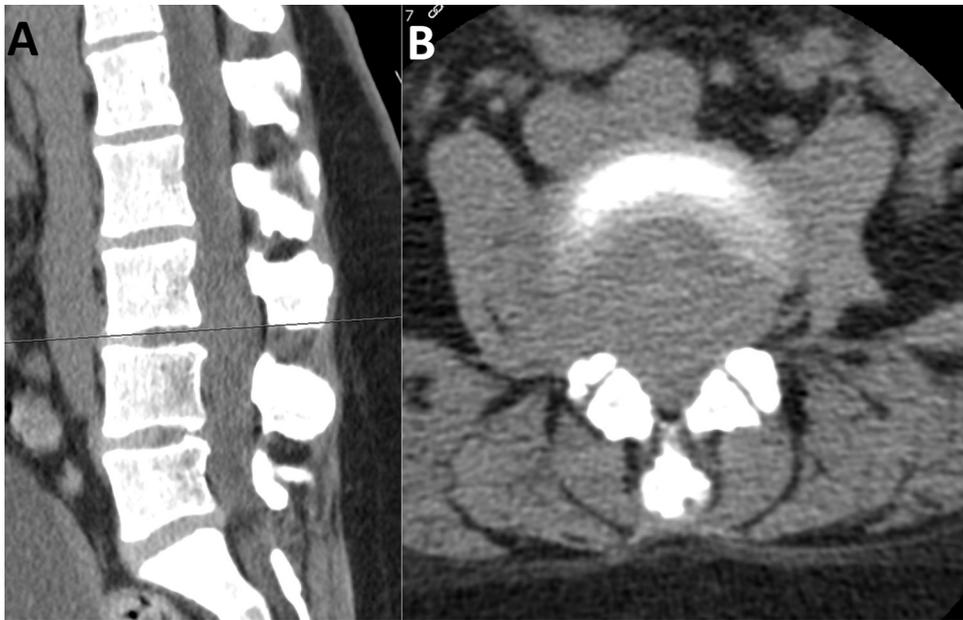


Fig. 1. Preoperative CT images. The lesion is barely visible and there is no bone destruction. A. Sagittal view, soft tissue windowing. B. Axial view, soft tissue windowing.

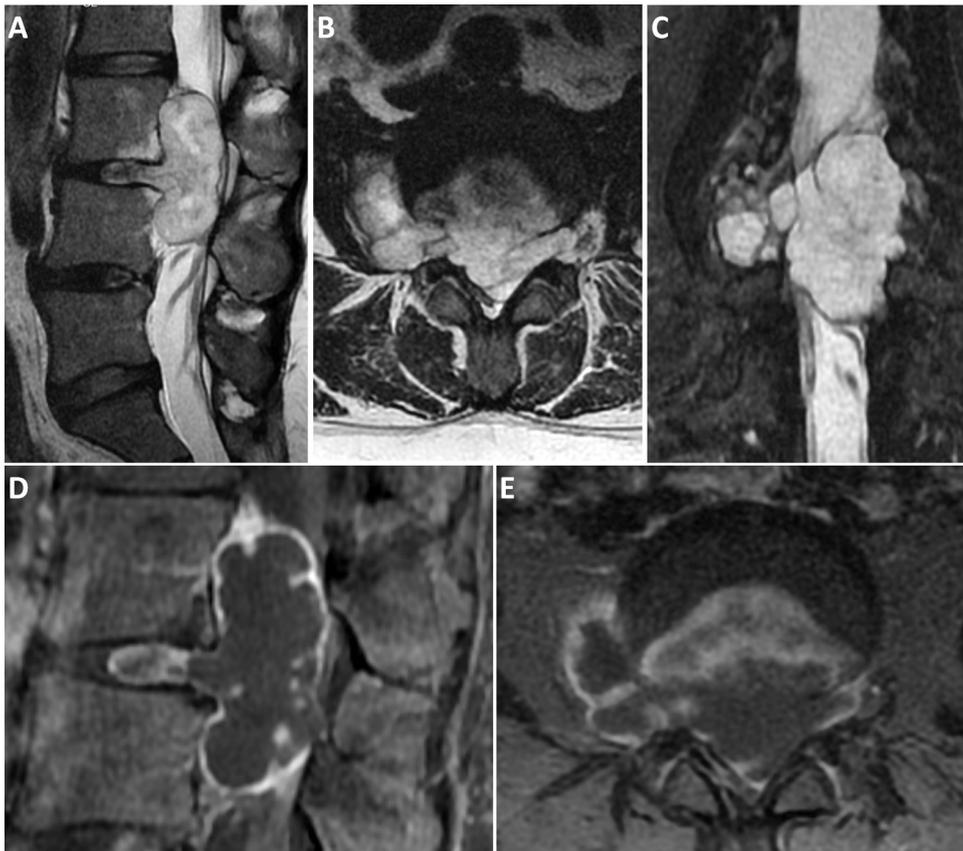


Fig. 2. Preoperative MRI images. A. Sagittal T2 image displaying a 52-mm high anterior epidural lesion originating from the L3-L4 disc, severely compressing the roots of the cauda equina. B. Axial T2 frFSE showing the lateral extension of the lesion into the psoas. C. STIR coronal view. D. Sagittal T1 with gadolinium enhancement showing a peripheral pattern of enhancement. E. Axial T1 FSE IV.

3.3. Histopathologic interpretation

The fragments consisted of nodules of well-differentiated cartilage without any evidence of bone or synovial tissue entrapped or surrounding the cartilaginous nodules. The matrix was hyaline

without myxoid alteration. Cellularity was low to moderate, with chondrocytes arranged in clusters, as commonly seen in synovial chondromatosis. Nuclei were enlarged, and binucleated cells were not infrequent. No necrosis or spindling was seen (Fig. 3). A chordoma was excluded. Tumor cells were strongly positive for S100

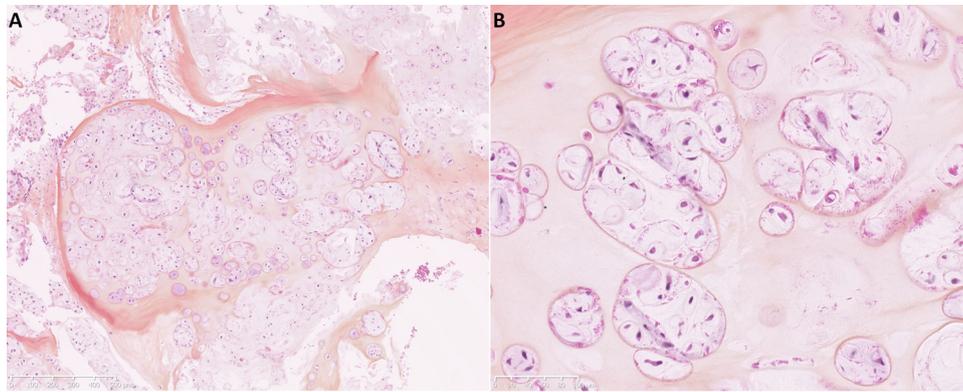


Fig. 3. Histopathologic microscopic views of the lesion. A. Hyaline cartilage nodules (Hematoxylin and Eosin, original magnification $\times 25$). B. Clusters of moderately atypical chondrocytes (Hematoxylin and Eosin, original magnification $\times 200$).

antibody, as expected in a well-differentiated cartilaginous tumor. After collegial review with the French Group of Bone Pathology, a reactive process was ruled out. The two hypotheses were: low-grade chondrosarcoma, or synovial chondromatosis.

3.4. Second surgical procedure and complications

6.3 months after the initial surgery, complete L2-L5 bilateral laminectomy with internal fixation was performed, followed by subtotal resection including the tumor invading the muscles. The same closure was made, with insertion of an external lumbar drain as the anterior wall of dura mater was torn apart. Microscopic examination of the tumor was unchanged but interpretation remained in doubt.

CSF leakage through the lumbar wound continued despite medical measures. Ten days later, redo surgery found no clear dural tear. Muscle, fascia, fat and glue were introduced in each lateral groove of the thecal sac and, a new external lumbar drain was inserted. CSF leakage decreased, but the patient developed *Escherichia coli* infection of the surgical site. Surgical lavage was performed and the septic syndrome lessened under appropriate antibiotherapy. The metalwork was changed 27 days after implantation. Finally, only a ventriculo-peritoneal shunt managed to stop the CSF leakage.

4. Results and progression

The patient was transferred to a rehabilitation center 60 days after the second hospital admission. After multiple reviews, definitive histopathologic diagnosis of low-grade chondrosarcoma of the disc was made. The case was discussed in a multi-disciplinary sarcoma and rare bone tumor team meeting. Radio- or chemotherapy was not indicated, but rather simple clinical and a radiological surveillance.

Two years after initial presentation, at the time of writing, the patient could walk independently with the aid of one crutch. Clinical examination showed significant right distal L5 and S1 paresis with reduced sensation, but no back pain or sciatica. Despite the metallic artifacts, there was no obvious tumor residue or recurrence on the last CT and MRI scans.

5. Discussion

The most common cause of sciatica is intervertebral disc herniation; thus, obscure causes are rarely considered in differential diagnosis.

Suspicion of acute cauda equina syndrome can easily be confirmed by MRI. Promptly operated on, outcome is often favorable. However, slow-growing tumoral processes, as in the present case,

may be more challenging to treat. Unexpectedly, the lumbar CT scan was not informative, which excluded tumor involving the bone, such as osteosarcoma. Intraoperatively, the tumor appeared to spread out from the intervertebral disc, with no primary facet joint, ligament or bone involvement. Size, location and local invasiveness of the dura mater initially hindered complete resection. In case discussion, a diagnosis low-grade chondrosarcoma of discal origin was suggested. However, this entity is not referenced in the World Health Organization (WHO) classification of tumors of soft tissue and bone (4th edition) [1,2]. It was decided to perform a second surgery to improve thecal sac decompression. En bloc resection with good oncologic margins was estimated as too risky and likely not feasible for this tumor displaying no formal aggressive histological features. This complex procedure was not attempted in first line, as it was considered more urgent to decompress the cauda equina and get a diagnosis of this peculiar looking lesion rather than to achieve full resection. Unfortunately, after the second procedure, the CSF continued leaking despite all measures. A ventriculo-peritoneal shunt finally managed to stop the leak [3,4]. After 6 surgical procedures in 7.5 months and intensive rehabilitation, the patient's clinical state was stabilized at the time of writing. Extrapolating recommendations for chondrosarcoma of the osseous spine, clinical and radiological surveillance was chosen, as low-grade chondrosarcoma is resistant to radiation therapy [5]. Due to artifacts generated by the metalwork, postoperative MRIs were not very informative.

Benign or malignant neoplasms involving the backbone are common, in contrast to those involving the intervertebral discs. Tumors arising from the disc are exceedingly rare. Moreover, despite the frequency of spinal metastases, the intervertebral disc is usually spared and conserves normal structure even after extensive collapse of the backbone body due to neoplastic tissue [6].

We performed a search of the medical literature, with no language restriction, for tumor of discal origin and differential diagnoses. Only a handful of reports describe very unusual and odd entities: e.g. giant intrapelvic malignant peripheral sciatic nerve sheath tumor, intradural ganglioneuroma, retroperitoneal liposarcoma or primary Ewing's sarcoma presenting as sciatica and/or disc herniation. None, however, resembled the present observation [7–12]. On the contrary, reports of migrated intervertebral disc fragments mimicking extra- or intradural spinal tumor are far more common, making this the main differential diagnosis [13].

Chondrosarcoma is a common primary slow-growing heterogeneous group of malignant bone tumors characterized by the formation of cartilaginous neoplastic tissue with varying morphologic features and clinical behavior. It is classified in 3 groups according to histopathologic characteristics such as mitosis rate: low-grade (grade 1), or grade 2 or 3. In the latest WHO classification,

low-grade chondrosarcoma was included under the “atypical cartilaginous tumor” subclass, to highlight its extremely heterogeneous aspect and indolent behavior with very low metastatic potential [14]. Around 10% of chondrosarcomas arise from the mobile spine, but the lumbar region is infrequently affected [15]. They generally present with pain in the area of the lesion and a palpable mass, unlike in the present observation. The present radiological features were unlike reported cases of spinal chondrosarcoma [16]. It rarely grows inside the spinal canal and almost always displays bone involvement [17]. Spinal chondrosarcoma is prone to local recurrence, despite being low-grade [16,18].

A review of the “disc chondrosarcoma” literature retrieved only one reference, a description of a chondrosarcoma of the temporomandibular joint arising solely from the disc itself [19].

Searching the literature, we reviewed many MRI images without finding any radiological aspect similar to that of the tumor reported here. We would ask anyone who has encountered such tumor with such an aspect to kindly contact us.

6. Conclusion

We report a very unusual and therapeutically challenging spinal tumor diagnosed as low-grade chondrosarcoma of discal origin, an entity never previously described.

Compliance with ethical standards

This study was conducted in accordance with the ethical standards of the Helsinki Declaration (2008). The patient gave her full informed consent for the publication of this report.

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Disclosure of interest

The authors declare that they have no competing interest.

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at <https://doi.org/10.1016/j.neuchi.2019.04.001>.

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