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Low-Grade Lumbar Chondrosarcoma Extending From L2 to S1 With Paraspinal Involvement: A Case Report

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Abstract

Chondrosarcoma (CS) is a rare tumor condition of varying grades and with variable biological behaviors, capable of producing cartilaginous material; these tumors can develop most frequently in the pelvis, skull base, and axial skeleton, being the second most common bone tumor. These neoplasms encompass a heterogeneous spectrum ranging from low-grade indolent lesions to aggressive neoplasms with high metastatic potential. Its high degree of development consists of multiple genetic instability events (COL2A1, ACAN, and COMP) and cellular components such as fibroblasts that favor the marked increase in chondroid material. Due to its variable growth rate, the clinical presentation of chondrosarcoma is usually insidious, which sometimes hinders an early diagnosis. In the present article, a patient is presented with a tumor and peripheral nervous symptoms that led to a surgical procedure in which a diagnosis of grade 1 chondrosarcoma (CS1) was made.

Categories: Pathology, Oncology, Neurosurgery
Keywords: atypical central cartilaginous neoplasm, chondrosarcoma, grade 1 chondrosarcoma, neoplasms, spine, tumor

Introduction

Cartilaginous tumors are a heterogeneity of neoplasms that generate an abundant hyaline cartilage matrix [1].

The yearly occurrence of chondrosarcoma (CS) is approximately 2-4 per million people per year, often affecting adults with an average age of 50-60, with a slight male predominance [2,3]. Within the osseous spine, chondrosarcoma has a predilection for the thoracic vertebrae but can arise anywhere along the length from the cervical spine to the sacrum [2].

They can be classified as either primary or secondary. Primary chondrosarcomas, which arise de novo, are the third most common primary malignant tumors of the bone after myelomas and osteosarcomas and account for 20%-27% of all primary malignant bone tumors [1,2].

Chondrosarcomas can also be classified based on the osseous location in which they arise, namely, central (within the intramedullary cavity), peripheral (within the cartilage cap of a pre-existing osteochondroma), or periosteal (juxtacortical; on the surface of the bone) [2,4].

The 2020 World Health Organization (WHO) classification categorizes chondrosarcomas into eight subtypes: central conventional (grade 1 versus 2-3), secondary peripheral (grade 1 versus 2-3), periosteal, dedifferentiated, mesenchymal, and clear cell [1,2].

The 2020 WHO classification categorizes chondrogenic bone tumors as benign, intermediate (locally aggressive), or malignant [2,3].

The biological behavior of chondrosarcomas is graded as 1-3 based on nuclear size, staining pattern (hyperchromasia), mitotic activity, and cellularity degree [2].

Another type of chondrosarcoma is the atypical cartilaginous tumor (ACT), which was first introduced in the 2013 WHO classification, also defined as grade 1; it is a hyaline cartilage-producing neoplasm, located in the appendicular skeleton [1]; meanwhile, grade 1 chondrosarcomas (CS1s) are designated for the axial skeleton (ranging from the pelvis, to the scapula, to the skull base). Although CS1s can be located at any level of the spinal segments, they have a predilection for the C2 vertebra and are more frequent than those of the appendicular skeleton [1,5,6].

Chondrosarcoma is the second most common nonlymphoproliferative primary malignant tumor of the spine in adults [5]. They encompass an immunological profile that varies depending on the aggressiveness of the disease: noninvasive low-grade chondrosarcomas are usually surrounded by CD8+ T cells, whereas invasive high-grade cases predominantly present an infiltration of M2 macrophages, in which microenvironment cells such as tumor-associated endothelial cells (TA-ECs) and fibroblasts also participate, contributing substantially to the malignancy of CS [7].

Various studies have been conducted to analyze which spinal segment is most frequently affected. In a publication by Giannini et al., the authors report on a cohort study of 67 patients diagnosed with low-grade chondrosarcoma (CS) of the spine. The majority of the tumors originated in the thoracic spine (76%), followed by the lumbar (15%) and cervical (9%) regions [5]. Although chondrosarcomas can be located at any level, the thoracic segments of the spine are the most commonly affected [8].

Clinically, it typically presents as a hard, fixed, and slow-growing mass, with the most common symptom of spinal chondrosarcoma being back or lumbar region pain, as was the case here, which is usually accompanied by neurological signs related to the compression of the spinal cord or nerve roots (such as radiculopathy, paresthesias, and paresis), and it is predominantly diagnosed in patients between 20 and 40 years of age [9,10].

Radiographic findings often suggest the diagnosis of chondrosarcoma because of the identification of typical "ring-and-arc" chondroid matrix mineralization (representing the enchondral ossification) and aggressive features of deep endosteal scalloping and soft-tissue extension. These latter features are usually best assessed, as is lesion staging, with computed tomography (CT) or magnetic resonance (MR) imaging. CT is optimal to detect the matrix mineralization, particularly when it is subtle or when the lesion is located in anatomically complex areas [10].

The extension of a mass from the extracanal space into the spinal canal is a feature observed mainly in neurogenic tumors (such as schwannomas or neurofibromas), making it an unusual finding in non-neurogenic tumors such as chondrosarcoma [8], and the external expansion of the tumor through the posterior elements of the spine allows for the palpation of a mass in the back or neck [11-13].

Histological grading is merely one indicator that can predict the tumor's biological behavior. Prognosis is also linked to therapeutic management [14].

The en bloc resection is associated with superior local control compared to subtotal resection, as prior studies have demonstrated an increased risk of locoregional recurrence with intralesional tumor excision; definitive en bloc resections are highly technical procedures performed at select centers, resulting in a reduced local recurrence rate and improved long-term survival [12,15,16].

Spine stereotactic radiosurgery (SSRS) with dose escalation has demonstrated improvements in the local control of chordoma or chondrosarcoma (histologies considered to be the most radioresistant types affecting the spine) compared with conventional palliative fractionation and may hold particular importance as a low-morbidity alternative to surgery in selected cases [15]. Chemotherapeutic agents have not proved to affect the outcome at all in spinal chondrosarcoma, and their role is limited [16].

Presented below is a rare case of lumbar chondrosarcoma, characterized by an insidious clinical presentation and a prolonged course, which was treated via surgical resection and instrumentation.

Case Presentation

A 57-year-old female patient began seven years prior to her hospital admission with low back pain, mild, and a numeric rating scale (NRS) of 1/10, intermittent, muscular type, and oppressive, which subsided spontaneously without the need for analgesics [17]. The pain increased gradually over the years, until one year prior to her surgery, when it reached an NRS of 8/10, constant, which subsided partially with the use of analgesics (she took up to three). In addition, she had volume enlargement in the right center-lateral lumbar area, well-localized, with well-defined borders, indurated and immobile, and no evidence of erythema. The oppressive characteristics are combined with neuritic-type pain in the right L2 and L3 radicular pathways. X-rays, spine tomography, and magnetic resonance imaging (MRI) demonstrate a solid tumor lesion involving soft tissues and the L2 and L3 neural foramina (Figure *1A*-*1E*).

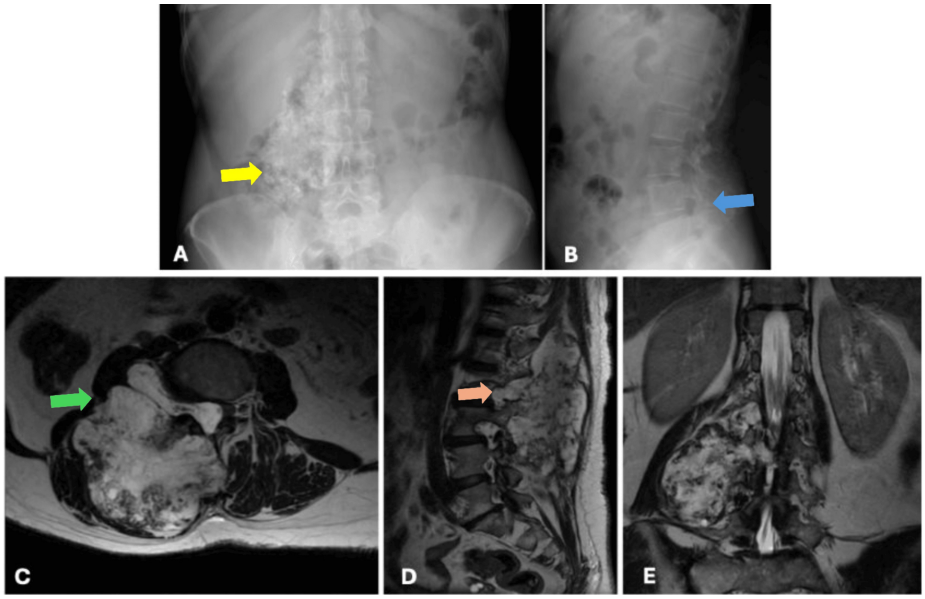


FIGURE 1: Preoperative studies.

(A and B) Preoperative X-rays showing a radiopaque area extending from vertebral levels L2 to S1 (yellow arrow), obscuring the spinous processes of L2-L5 (blue arrow). (C-E) T1-weighted MRI of the lumbosacral spine. Note the heterogeneous characteristics, predominantly hyperintense and isointense, and the displacement of the psoas muscle, lumbar musculature, and kidney (green arrow). This results in stenosis of the L1-L2, L2-L3, and L3-L4 neuroforamina (orange arrow), as well as secondary lumbar spinal stenosis at the same levels.

MRI: magnetic resonance imaging

Surgical procedure

In the prone position, a midline incision is made, and the tumor border deep to the subcutaneous cellular tissue is located. It is dissected en bloc, detaching it from its deep face, as well as from the vertebral insertion. Psoas and renal borders were manually dissected (Figure *2A*-*2H*).

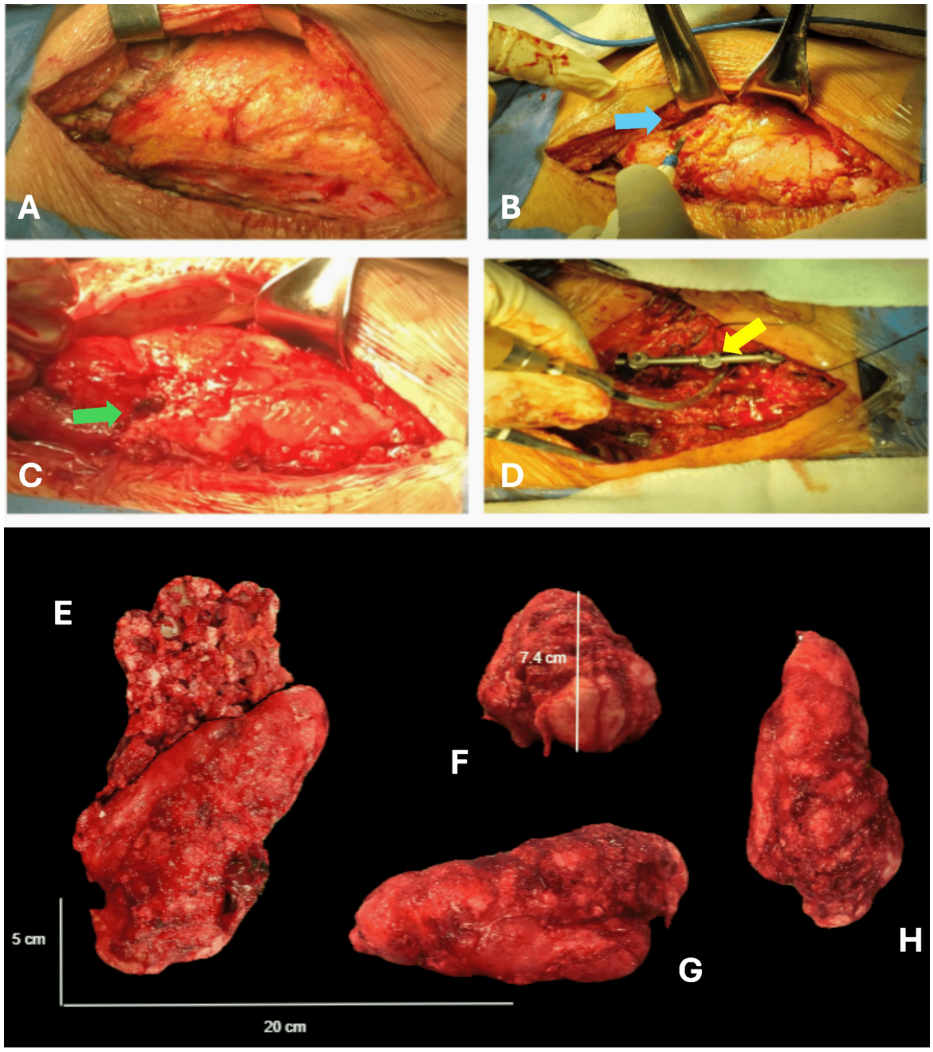


FIGURE 2: Intraoperative images.

(A and B) Exposure of the muscle and tumor and tumor dissection (blue arrow). (C and D) Tumor coagulation (green arrow) and the placement of screws to improve vertebral stability (yellow arrow). (E-H) Biological specimen in superior, lateral, posterior, and anterior views.

By means of microscopy, foraminal and affected lumbar canal decompression was completed. L2, L3, and L4 transpedicular instrumentation with rods and screws is shown in Figure *3A*-*3F*.

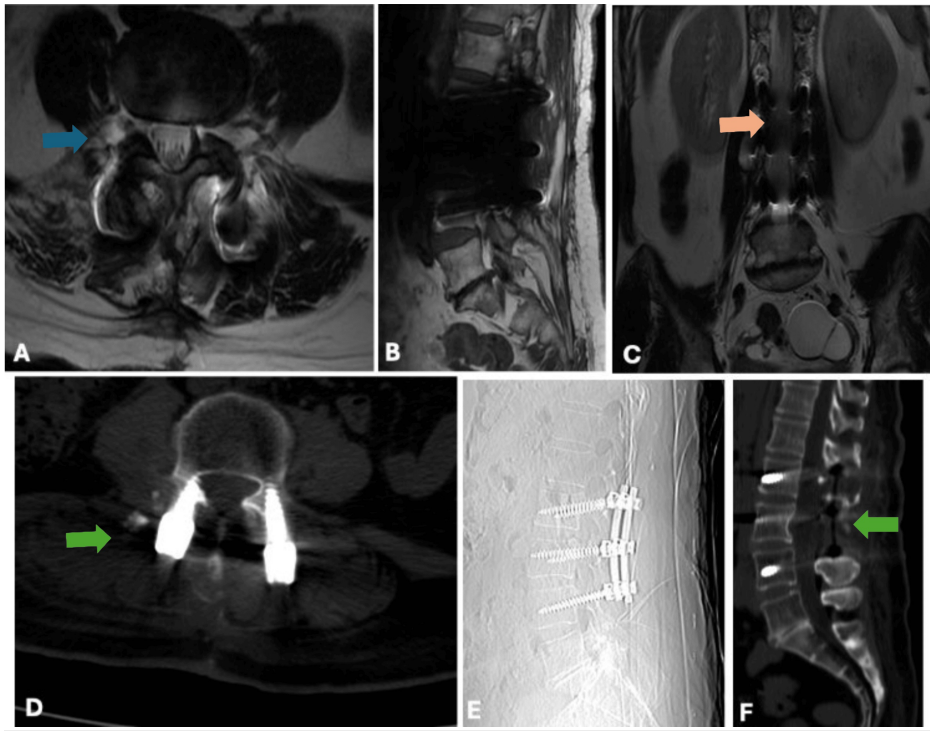


FIGURE 3: Postoperative images.

(A-C) T2-weighted MRI of the lumbosacral spine. Note the decompression of the lumbar canal (orange arrow), the inferior displacement of the right kidney, and the return of the right psoas muscle to its normal position (blue arrow). (D-F) Plain CT scan of the lumbosacral spine showing the resolution of the case with L2-L4 transpedicular screws and rods and the resection of the chondrosarcomatous lesion (green arrows).

CT, computed tomography; MRI, magnetic resonance imaging

The pain decreased to an NRS of 1/10. There were no motor sequelae. No complications or need for surgical intervention was documented. The pathology result, with folio number RL19-2359, was reported as a well-differentiated conventional-type chondrosarcoma, grade 1 without the presence of necrosis.

Protocol and follow-up studies were conducted, showing no evidence of metastasis to date (Figure *4A*-*4C*).

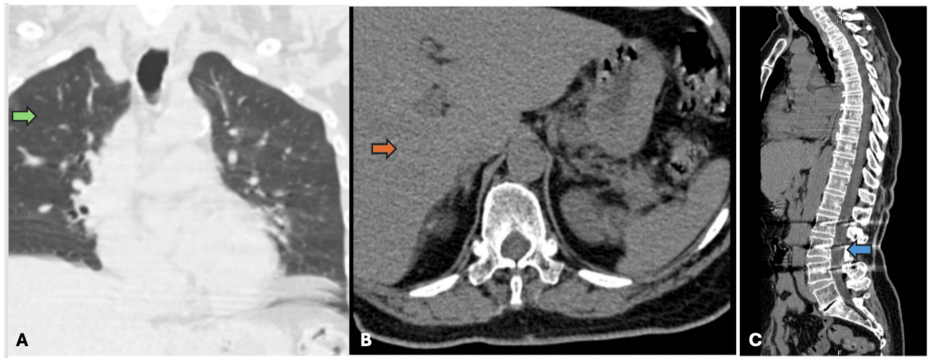


FIGURE 4: Systemic studies to rule out metastasis.

(A and B) Thoracoabdominal CT; expanded lung fields are observed with homogeneous lung parenchyma of normal attenuation; no nodules, consolidations, or space-occupying lesions are seen in the pleural space (green arrow); the liver presents normal morphology, contours, and size and is homogeneous, without structural abnormalities (orange arrow). (C) Non-contrast CT of the cervicothoracic and lumbosacral spine showing normal vertebral bodies and intraspinal spaces, with postsurgical changes secondary to the resection of the chondrosarcomatous lesion and the placement of transpedicular screws and rods at levels L2-L4 (blue arrow).

CT: computed tomography

The patient was referred to medical oncology but did not require adjuvant treatment. Annual follow-up was maintained; follow-up now extends to five years, with no evidence of lesion recurrence on contrast-enhanced MRI (Figure *5A*-*5C*).

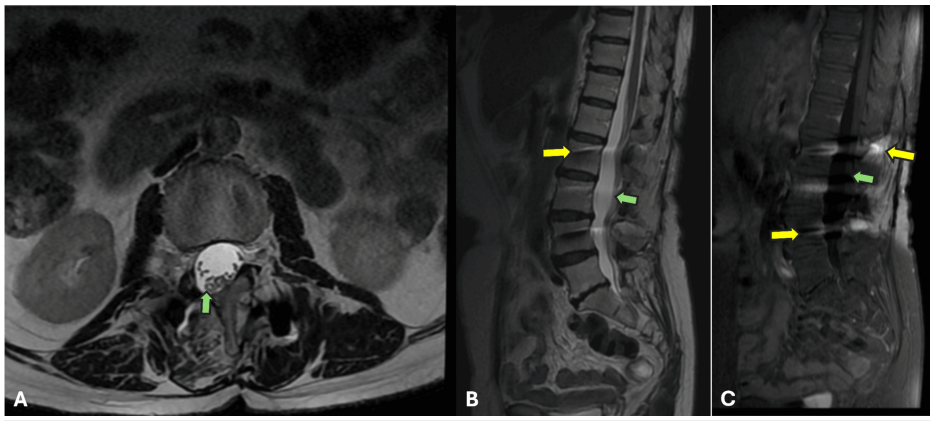


FIGURE 5: Five-year follow-up images.

(A and B) T2-weighted lumbosacral spine MRI. Note the changes secondary to surgery, showing adequate decompression of the lumbar canal (green arrows). (C) Sagittal view with gadolinium; no enhancement is observed upon contrast administration to suggest recurrence of the chondrosarcomatous lesion, with imaging artifact due to osteosynthesis material (yellow arrows).

Discussion

The distinction of ACT/CS1 from other grades of CS lies in the mitosis and nuclear pleomorphism present in other grades [1].

Unlike chondrosarcomas, enchondromas are benign hyaline cartilage tumors originating from the medullary cavity of the bone [1].

These lesions are also divided into primary lesions, "de novo" CS, or secondary lesions, subsequent to a pre-existing benign lesion such as an enchondroma [1,2,6]. Furthermore, the location of lesion formation distinguishes a central (intraosseous) lesion from a peripheral (exostosis) one [1,2,6].

ACT/CS1s show a lobular growth pattern, low cellularity, and a predominantly hyaline cartilaginous matrix, cellular binucleation with the absence of mitoses, and the two criteria to differentiate from an enchondroma are as follows: the lamellar entrapment of the host bone and mucoid or myxoid changes of the surrounding matrix [1].

The high development of cartilaginous material can generate a larger size that invades the walls of the spinal canal and compromises spinal stability, reaching very large dimensions that even span the spaces of the erector spinae and quadratus lumborum muscles. After completing the extraction procedure of the tumor mass, vertebral compromise and its stability must be considered [11,13,14].

Surgery is of critical importance when treating spinal chondrosarcomas and should aim at preserving or even improving functionality, relieving pain, and controlling local tumor recurrence, promising a prolonged survival. The spectrum of oncologically established surgical procedures applicable to the spinal column varies from the most complex en bloc resection to the simplest one implying a piecemeal removal of the tumor (curettage) [15,16].

Besides the histological grade of the tumor, the prognosis depends on the possibility of performing en bloc excision with appropriate oncologic margins. A successful operation, in terms of complete tumor excision with disease-free margins, is a major independent prognostic factor for a favorable course of the disease, affecting both local tumor control and patient survival critically [16].

The patient's age, the extent of the disease at the time of diagnosis, and the grade of the lesion significantly influence the prognosis, as does the chosen treatment, whether surgical or radiation-based.

Conclusions

Chondrosarcomas are rare lesions whose main feature is the production of cartilaginous material. They can vary in size, location, and consequently their clinical characteristics. In the present case, the lesion was

causing a narrow lumbar canal, clinically characterized by axial pain, as well as radiculopathy due to its foraminal and extra-spinal extension. These are grade 1 lesions, but their behavior tends to be locally invasive, causing the compression of adjacent structures.

Surgical treatment is recommended to improve symptoms and prognosis, depending on the tumor's extent and the associated comorbidities in each case. In this case, adequate decompression and tumor resection were achieved, resulting in immediate clinical improvement.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

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Disclosures

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