

# Large Chondrosarcoma of the Lumbar Spine – A Rare yet Important Cause of Lower Back Pain

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## ABSTRACT

*We report a case of a large chondrosarcoma of an L4 vertebral body causing iliac vein thrombosis. The slow-growing tumor eluded definitive diagnosis early in its development since the main symptom it caused was only lower back pain. Five years after onset of the disease, the patient presented with fever, tenderness and swelling in the leg, the tumor was diagnosed and found to be exerting a mass effect causing further pain and compressing the left common iliac vein. Due to inoperability of the tumor, a multidisciplinary surgical approach was used to resect the majority of the tumor as a palliative measure and rid the patient of her symptoms. Due to the chemoresistance and relative radioresistance of these tumors, prompt full surgical resection before the tumor invades vital structures remains the mainstay of successful treatment of chondrosarcoma of the spine.*

**Key words:** chondrosarcoma, spine, iliac vein, thrombosis, case report, surgery

## Introduction

Chondrosarcoma belongs to the family of malignant tumors (though a rare neoplasm on the whole, it stands third among malignant primary bone tumors, after osteosarcoma and Ewing's sarcoma)<sup>1,2</sup>.

The maximum incidence of chondrosarcoma is found in patients 30 to 70 years of age<sup>3,4</sup>. Rarely does chondrosarcoma occur before the age of 20 years or over 70, and only exceptionally is it found before puberty<sup>1,5-7</sup>. Chondrosarcoma occurs 3–12% of the time in the spine, and it represents 7–12% of all spinal neoplasms<sup>2,8,9</sup>.

Due to the rarity of primary spinal chondrosarcoma, there are only few descriptions of large series of patients with spinal chondrosarcoma in the literature<sup>6,9</sup>. Histologic grading of chondrosarcoma ranges from 1 to 3 (in some reports 0 to 3)<sup>2,3,10,11</sup>. Though a look into the biological behavior of chondrosarcoma is beyond the scope of this paper, they have been shown to display a wide variety of molecular compositions between specimens, thus implying many genetically different species of tumour<sup>10</sup>. Radiographically, though different subtypes of the tumor have different characteristics, chondrosarcoma is typically associated with a large area of bone destruction and a soft tissue mass having calcifications within<sup>8</sup>. Magnetic

resonance is the best imaging modality for evaluation of the lesion and potential cord compression<sup>8</sup>. Due to the slow growth of this neoplasm, patients often will present with a rather advanced form of the disease. The most common presenting symptom is localized pain, followed by a palpable mass<sup>9</sup>. Neurological disturbances have been described in up to 50% of patients at presentation<sup>2,9</sup>. Because of its slow growth, local recurrence and metastases may occur more than 10 years after removal of the tumor. These tumors are resistant to most protocols of radiation therapy and chemotherapy, though there is some evidence that radiation therapy may be of use in inoperable cases or after intralesional resection<sup>9,12</sup>. The surgical management for chondrosarcoma is clearly established and commonly accepted. The prognosis depends on the histologic grade of malignancy and on feasibility of performing en-bloc excision with appropriate oncologic margins<sup>4,7</sup>.

## Case Report

In 1994, a 46 year old female presented to another institution with pain in her lumbar region spreading to

both lower limbs. After plain radiographs and computerized tomography (CT) scan of lumbar spine (Figures 1 and 2), status post pathological compressive fracture of L4 or metastatic tumor were suspected as the most likely causes. A thorough diagnostic workup did not find signs of a primary tumor.



Fig. 1. Lateral view plain film radiograph at initial presentation. At this point the tumor is still localized to the boundaries of the vertebral body and clinically only caused lower back pain.



Fig. 2. Computerized tomography scan through the L4 body giving a typical appearance of a burst fracture of a vertebral body.

Five years later, fever, swelling, tenderness and pain in her left lower limb developed. Scintigraphy showed complete left common iliac vein thrombosis. By that time there was a palpable tumor in her lower abdomen, which was attributed to a coincidentally present myomatous uterus. The symptoms persisted after hysterectomy. Follow-up CT of the lumbar spine showed tumor expansion (10 x 15 x 15 cm) to the lower pelvic region (Figure 3).

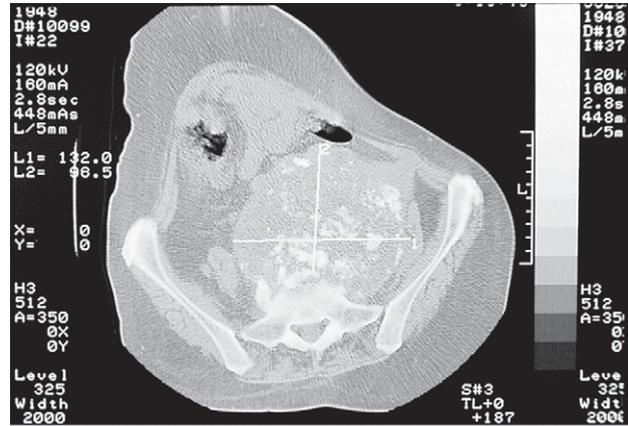


Fig. 3. Computerized tomography scan at presentation to our institution, 5 years after initial presentation of lower back pain. Note the dimensions of the tumor – 132mm x 96.5mm. At this point the tumor was exhibiting a mass effect on the iliac veins.

The patient then came to our department and a biopsy was performed confirming the diagnosis of a low grade classic chondrosarcoma.

Angiographic examination confirmed complete left iliac vein thrombosis and almost complete obstruction of the right iliac vein and development of large collateral veins (Figure 4). These characteristics made complete excision of the tumor impossible.



Fig. 4. Venography of the iliac veins. Note the severe stenosis of the right common iliac vein and the complete absence of contrast in the left iliac veins.

Due to the size of the tumor and the fact that it compromised such large blood vessels and the ureters, the surgical team consisted of a vascular surgeon, urologist, and an orthopedist.

The most demanding part of the surgery was the ligation of the very large collateral veins around the tumor,

which was done from an anterior approach. The second stage of the surgery was a partially anterior and partially posterior tumor reduction and fixation of adjacent vertebrae.

After this paliative procedure, the patient was free of pain and swelling in her lower limbs. Two years after surgery she suddenly died of a massive pulmonary embolism, which was not linked to recurrence of local disease upon autopsy.

## Discussion

The mainstay of treatment for chondrosarcoma is surgical excision, but options are limited or dictated by the tumor's proximity to vital structures as well as the risk of jeopardizing stability<sup>6,13</sup>. Most authors agree that a wide resection, that is, removing the tumor en bloc with a thin layer of healthy surrounding tissue and without damaging or exposing the interior of the tumour, is key in minimizing recurrence years later<sup>2,3,14,15</sup>. Prognosis is related to histologic grading and management<sup>7</sup>. Because this condition is typically resistant to the known protocols of chemotherapy and is also relatively radioreistant, the role of surgical management is preeminent. It is important to note that in a study of chondrosarcomata, the follow up period should be longer than for other primary tumors as it is often reported to recur 10 years upon surgical intervention<sup>4</sup>. Lower grade chondrosarcomata have been noted to dedifferentiate into higher grades upon recurrence, though this phenomenon is more uncommon in young patients<sup>1,9</sup>. Higher grade tumors naturally have a higher chance of metastasizing (the lungs being a particularly common site of distant seeding)<sup>6,12</sup>.

In this case report, our patient, like many others, in large series had a very long history of back pain that had not been adequately investigated. Afterwards, she developed symptoms related to the compression of large vessels which were associated with a myomatous uterus. After the hysterectomy, symptoms persisted and a follow-up

CT of the lumbar spine reveals a large tumor mass spreading into the pelvis, compromising the veins and ureters. Because of the inoperable nature of the tumor, the specialist team decided to perform a partial resection of the tumor to allow for decompression of the blood vessels and ureters. However, such intralesional surgical resection is known to have a higher rate of recurrence years later than wide or radical excision<sup>4</sup>.

Hemicorporectomy was even discussed as an alternative procedure. Hemicorporectomy (translumbar amputation) is a severely mutilating procedure which can be performed for malignant conditions not otherwise amenable to surgical resection. The patient refused to undergo such a procedure. Unfortunately, the patient suddenly died 2 years after operation from an apparent massive pulmonary embolism.

## Conclusion

Spinal tumors must be considered in the differential diagnosis of back pain untracable to more common causes. Though rarely encountered in clinical practice, such tumors can be treated much more effectively if uncovered early on, using »red flag« signs of spinal malignancy as a guide to clinical reasoning (are: age >50 years, no improvement in symptoms after one month, insidious onset, a previous history of cancer, no relief with bed rest, unexplained weight loss, fever, thoracic pain, or being systematically unwell<sup>16</sup>) before having had a chance to grow and spread to local neurovascular structures or organs. The lower the grade and the smaller the discovered tumour, the better will the prognosis be on the whole. Total resection offers the best chance for a prolonged disease-free interval in patients with spinal chondrosarcoma though there may be a place for adjuvant radiotherapy when complete resection is not possible, particularly in high grade tumors. Recent advances in spinal stabilization and in surgical approaches to the spine now allow the surgeon to excise these tumors more completely, providing better prognosis for patients.

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## **VELIKI HONDROSARKOM SLABINSKE KRALJEŽNICE – RIJEDAK ALI OZBILJAN UZROK KRIŽOBOLJE**

### **S A Ž E T A K**

Prikazujemo slučaj opsežnog hondrosarkoma L4 kralješka koji je u bolesnici uzrokovao trombozu ilijakalnih vena. Ovaj sporo rastući tumor je izbjegao točno dijagnosticiranje u ranoj fazi razvoja jer je glavni simptom bio samo križobolja. Nekoliko godina nakon početka simptoma bolesnica prezentira sa vrućicom, bolnosti i oteklinom u nozi, tumor je dijagnosticiran i otkriveno je kako mehanički pritišče okolne strukture uključujući lijevu venu iliacu communis. Zbog inoperabilnosti ovog tumora, multidisciplinarni zahvat je učinjen kao palijativna mjera kako bi se simptomi suzbili. Radi otpornosti ove vrste tumora na kemoterapiju i radioterapiju, rano i opsežno kirurško liječenje prije nego li dođe do invazije okolnih struktura je standard liječenja hondrosarkoma kralježnice.