

Chondrosarcoma in the body of the C2 vertebral axis

Baris Ozoner, MD; Ahmet Kayhan, MD; Nil Comunoglu, MD; Seckin Aydin, MD; Galip Zihni Sanus, MD; Necmettin Tanriover, MD

ABSTRACT

Chondrosarcoma, a malignant bone tumor, is rarely encountered in the cervical spine. This article describes a patient whose neck pain and dysphagia were caused by an expansive, destructive lesion with calcification that was located in the body of the axis (C2 vertebra), the first time a chondrosarcoma has been reported in this location.

Keywords: chondrosarcoma, spine, cervical vertebra axis, endoscopy, transoral surgery, spinal neoplasm

CASE

A 39-year-old woman was referred to our clinical for neurosurgical consultation after imaging studies detected a mass in the C2 vertebra. The patient initially presented with neck pain and difficulty swallowing.

History The patient had experienced cervical pain for the past 10 years that had become worse after a fall about 1 year ago. The pain was localized to the back of her head and the upper cervical region. For the past 6 months, she also experienced difficulty swallowing, especially solid foods.

Physical examination Inspection and palpation of the head and neck were normal. The patient had moderate pain when her neck was forced to flexion. Her breathing was regular and unlabored. Neurologic and physical examination revealed only dysphagia. The patient had no history of chronic disease.

Diagnostic testing Laboratory testing revealed no abnormalities. Diagnostic interventions were made to examine

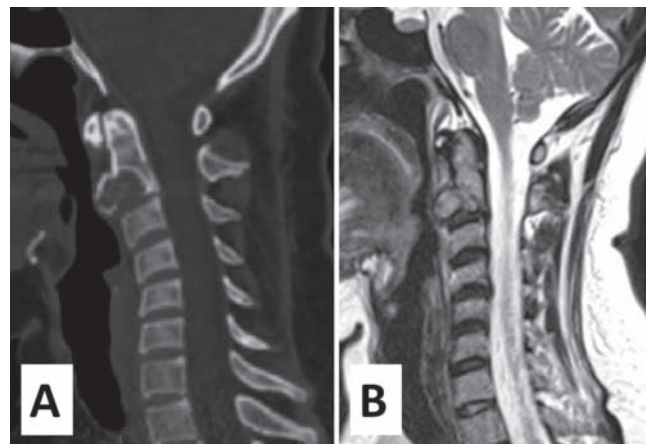


FIGURE 1. Preoperative images of the cervical vertebra: sagittal section CT (A) and T2-weighted image MRI (B)

the compression of the mass to pharyngeal and neural structures, and to evaluate cervical stability. Endoscopic examination of the oropharyngeal region showed a barely visible mild swelling along the posterior part of the pharynx that was devoid of desquamation or abrasion of the pharyngeal mucosa. Plain radiographs showed a destructive lesion in the C2 corpus. A cervical spine MRI showed an antero-inferior intracorporeal tumor that extended from the anterior margin of the lower C2 body and compressed the posterior part of the pharynx. The tumor dimensions were 18x10x9 mm; it was found to be hypointense in T1-weighted images and hyperintense in T2-weighted images with heterogenous enhancement after contrast administration (Figure 1). CT of the cervical spine revealed an exophytic mass in the antero-inferior part of C2, along with localized bony destruction and calcified regions in the mass (Figure 1).

Management Based on the patient's dysphagia and cervical pain symptoms, and the pharyngeal compression noted in radiologic and endoscopic examinations, the healthcare team decided to remove the mass via endoscopic transoral surgery.

With the patient under general anesthesia, a nasogastric (NG) tube and Boyle-Davis mouth gag divider were used for a scant jaw opening. The uvula was pulled upward

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Key points

- Solitary spinal chondrosarcomas are most frequently located in the thoracic spine.
- Chondrosarcomas predominantly arise in the posterior compartments of the spine.
- Chondrosarcoma rarely is isolated in the body of the cervical vertebra.
- This is the first case report of chondrosarcoma that confined in the body of the C2 vertebra (axis).

via feeding tube (Figure 2 and Video 1 at www.jaapa.com). A vertical midline incision on the pharynx and lateral retraction of muscles allowed visualization of the anterior part of the mass. The tumor was pearl-colored and solid and was removed using endoscopic curettes, punches, and a high-speed drill. After removal of the entire tumor, about 2 mm of healthy bone was drilled for the en-bloc resection. Following the resection, the muscle and mucosa were sutured. The patient was fed through an NG tube until postoperative day 5 to ensure healing of the pharyngeal mucosa. An early postoperative cervical spine MRI and thin-slice CT images were performed on postoperative day 1 to confirm complete removal of the tumor (Figure 3). The patient was mobilized with a cervical collar, the NG tube was removed on postoperative day 5, and the patient was discharged on postoperative day 10. Low-grade chondrosarcoma, with low cellularity and mild atypia in tumoral cells, was detected in pathological examinations of the surgical samples (Figure 4). Tumoral infiltration was observed in the host bone (Figure 5). No cervical instability and no recurrence was observed in the 8 months of follow-up.

DISCUSSION

Chondrosarcoma is the third most common primary malignant bone tumor after osteosarcoma and Ewing sarcoma and encompasses a heterogeneous group of transformed cells that are characterized by their ability to form cartilage.¹ The reported incidence of solitary spinal chondrosarcoma is less than 10% and tumors are most frequently located in the thoracic spine, followed by the lumbar spine.¹⁻³ Half of patients present with neurologic symptoms and a palpable mass and the most common symptom is pain.¹ Occasionally, chondrosarcoma may occur in the vertebral body of the spine (5%), and lesions predominantly arise in the posterior elements (40%), or in both anterior and posterior compartments (45%).¹

To the best of our knowledge, very few previously reported cases of chondrosarcoma were located in the cervical vertebra.⁴⁻⁶

In a case of chondrosarcoma located in the body of a cervical vertebra, the patient had a compression frac-

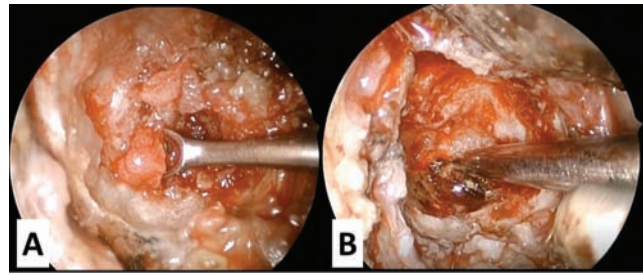


FIGURE 2. Intraoperative views of the mass located in the corpus of the C2 vertebra: using an endoscopic curette during tumor removal (A) and after total resection (B)

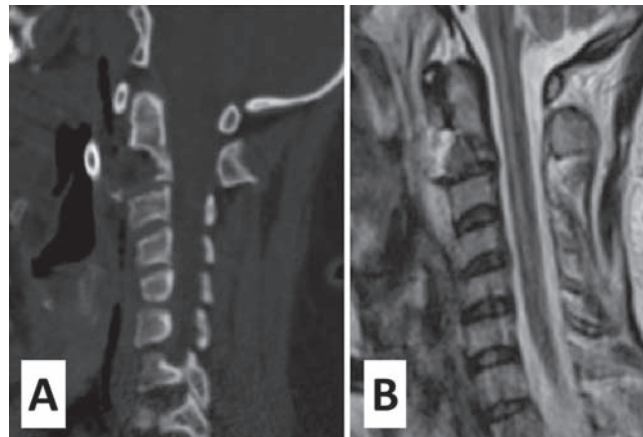


FIGURE 3. Postoperative images: sagittal CT (A) and sagittal T2-weighted MRI (B)

ture after a fall due to the lytic lesion in C7.⁷ Clear-cell chondrosarcoma was detected and treated with partial corpectomy.⁷

Our case is unique in regard to its location along the axial axis because the tumor was solely confined to body of the lower part of the C2 vertebra.

Chondrosarcomas have been classified into three grades; this grading is the best predictor of clinical behavior.⁸

- *Grade I* lesions are low-grade, less cellular, well-differentiated tumors that have a hyaline cartilage matrix and rarely metastasize.
- *Grade II* lesions are average-grade tumors with lesser matrix, but increased cellularity.
- *Grade III* lesions are high-grade tumors with high cellularity, mitoses, and anaplastic cells, 70% of patients with this tumor grade have metastases.⁸

In this patient's case, because histologic analysis of the tumor showed low cellularity and mild atypia (Figure 4) with tumoral infiltration of the host bone (Figure 5), it was pathologically scored as a grade I.

Chondrosarcomas are composed of a mineralized chondroid matrix and T2-weighted images of the tumor showed high signal intensity, which may be due to the high water

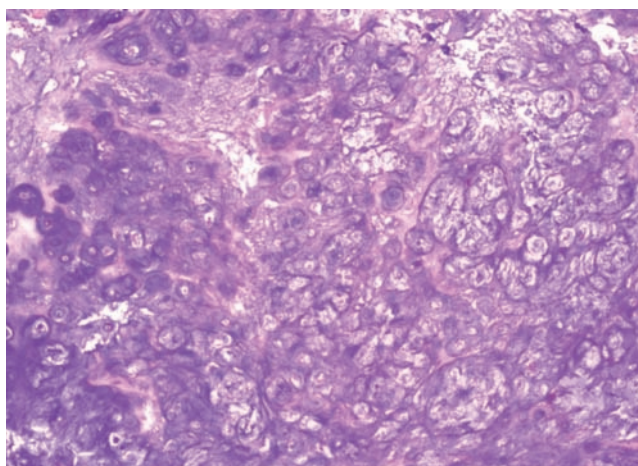


FIGURE 4. Low-grade chondrosarcoma at 100 times magnification, showing low cellularity and mild atypia in tumoral cells

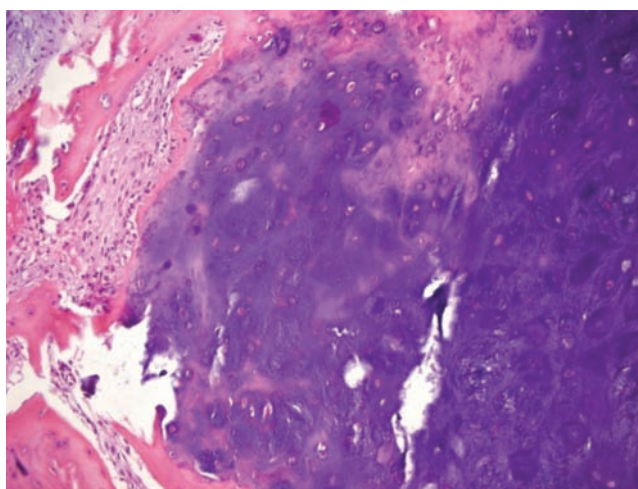


FIGURE 5. Tumoral infiltration to the host bone, at 200 times magnification

content of the hyaline cartilage.⁹ In contrast, calcified mineralization shows low signal intensity on all MRIs and a characteristic ring-and-arc pattern.⁹ However, calcified regions after matrix mineralization are more frequently detected with CT; in this patient's case, the calcified region was observed on MRI and CT. The pattern of contrast enhancement is mostly peripheral or septal in low-grade lesions, and characteristically nodular or has diffuse enhancement in high-grade lesions.¹⁰ The tumor in this patient's case showed peripheral enhancement with appropriate pathologic behavior.

Chemotherapy and radiotherapy usually are ineffective for spinal chondrosarcomas.¹ The en-bloc surgical removal (removing the tumor in a single piece encircled by a minor piece of healthy tissue) of the mass is the most successful treatment option.^{1,4,5} En-bloc resection with disease-free margins offers the best outcome for local tumor control,

with recurrence rates of 3% to 8%.^{1,11} In contrast, curettage procedures or intralesional removal have up to a 100% recurrence rates.^{1,12} In the case patient, a purely endoscopic transoral approach for gross total resection was used. Direct endoscopic visualization of the lesion provided sufficient operational view and ease of surgical maneuvers during surgery. Removing a small piece of nontumoral bone from the body of the axis using a high-speed drill ensured total removal of the lesion without destabilizing the cervical spine.

CONCLUSION

We believe that this case report is the first of chondrosarcoma in the lower body of the axis. Radiologically, the tumor had calcified regions, and histologically, it showed low cellularity and mild atypia with infiltration of the host bone, confirming the tumor was a low-grade chondrosarcoma.

Consider cervical spinal tumors in patients presenting with neck pain and swallowing disorders. Stabilize the patient's neck with a rigid collar to reduce the risk of dislocation, and monitor for signs and symptoms of tumor recurrence after surgery. **JAAPA**

REFERENCES

1. Katonis P, Alpantaki K, Michail K, et al. Spinal chondrosarcoma: a review. *Sarcoma*. 2011;2011:378957.
2. Riedel RF, Larrier N, Dodd L, et al. The clinical management of chondrosarcoma. *Curr Treat Options Oncol*. 2009;10(1-2):94-106.
3. Sundaresan N, Rosen G, Boriani S. Primary malignant tumors of the spine. *Orthop Clin North Am*. 2009;40(1):21-36.
4. Merchant S, Mohiyuddin SMA, Rudrappa S, et al. Cervical chondrosarcoma--rare malignancy: a case report. *Indian J Surg Oncol*. 2014;5(4):293-296.
5. Matsumoto Y, Takahashi Y, Harimaya K, et al. Dedifferentiated chondrosarcoma of the cervical spine: a case report. *World J Surg Oncol*. 2013;11:32.
6. Gietzen L, Pokorski P. Chondrosarcoma of the cervical spine. *JAAPA*. 2017;30(12):23-25.
7. Tessitore E, Burkhardt K, Payer M. Primary clear-cell chondrosarcoma of the cervical spine, case illustration. *J Neurosurg Spine*. 2006;4:424.
8. Evans HL, Ayala AG, Romsdahl MM. Prognostic factors in chondrosarcoma of bone: a clinicopathologic analysis with emphasis on histologic grading. *Cancer*. 1977;40(2):818-831.
9. Murphey MD, Walker EA, Wilson AJ, et al. From the archives of the AFIP: imaging of primary chondrosarcoma: radiologic-pathologic correlation. *Radiographics*. 2003;23(5):1245-1278.
10. Aoki J, Sone S, Fujioka F, et al. MR of enchondroma and chondrosarcoma: rings and arcs of Gd-DTPA enhancement. *J Comput Assist Tomogr*. 1991;15(6):1011-1016.
11. Boriani S, De Iure F, Bandiera S, et al. Chondrosarcoma of the mobile spine: report on 22 cases. *Spine (Phila Pa 1976)*. 2000;25(7):804-812.
12. Quiriny M, Gebhart M. Chondrosarcoma of the spine: a report of three cases and literature review. *Acta Orthop Belg*. 2008;74(6):885-890.