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Michael Nicolette
*Rowan University*, nicole57@rowan.edu

Swar Vimawala
*Cooper University Hospital*, vimawala-swar@cooperhealth.edu

Xinmin Zhang
*Cooper University Hospital*, Zhang-xinmin@cooperhealth.edu

Corey Mossop
*Cooper University Health Care*, Mossop-Corey@cooperhealth.edu

Brian Swendseid
*Cooper University Hospital*, swendseid-brian@cooperhealth.edu



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Case Reports and Case Series

# A Case of Cervical Chordoma

Michael Nicolette[1], Swar Vimawala, MD[2], Xinmin Zhang, MD[3], Corey M. Mossop, MD, FAANS[4], Brian Swendseid, MD[2]

[1] (Medical Student), Cooper Medical School of Rowan University, [2] Otolaryngology, Cooper University Health Care, [3] Pathology, Cooper University Health Care, [4] Neurosurgery, Cooper University Health Care

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A 48 year old male presented with neck pain and left arm pain that worsened with movement alongside left arm weakness, numbness, and tingling. The differential diagnosis for a patient with symptoms of a neck mass includes benign tumors like neurofibromas, malignant tumors like chordomas, and non-neoplastic conditions like cervical spondylitis. A magnetic resonance imaging (MRI) study of the cervical spine with and without contrast identified a T1 hypointense, T2 hyperintense, heterogeneously enhancing prevertebral mass with parapharyngeal extension. A direct laryngoscopy with biopsy was performed and revealed a paraspinal tumor. The patient's diagnosis of cervical chordoma was confirmed upon detection of Brachyury, a gene that encodes a transcription factor which promotes epithelial mesenchymal transition (EMT) in chordoma pathogenesis. Chordomas are slow-growing tumors located within the body's midline and they are associated with poorer outcomes because of neurovascular encasement at time of presentation. Chordomas are rare, with an incidence of 1 in 1,000,000. Approximately 6% of chordomas are located in the cervical spine. They are typically treated with surgery followed by radiation therapy. The patient underwent anterior resection for the prevertebral mass in the C2-C6 section of the cervical spinal cord. Surgery achieved subtotal resection and involved removal of the spinous processes of C3-C5 and reconstruction of the cervical spine with implants. The patient will be starting proton beam radiotherapy for his adjuvant treatment. Although rare, it is important to keep chordomas in the differential diagnosis when evaluating a patient with a neck mass.

## INTRODUCTION

Chordomas are invasive tumors of the midline and can be lethal without proper intervention. They are considered a rare disease with an incidence of 1 in 1,000,000. Although rare, chordomas are the most common primary cause of spinal tumors in adults.[1] Cervical chordomas constitute only 6% of all chordomas and are challenging to treat because of their proximity to important structures and late presentation.[1,2] Chordomas at any level of the spine can cause neurological symptoms such as radiculopathy by impinging on nerves that exit the spinal cord, and cervical chordomas specifically can cause dysphagia by compressing the esophagus.[2] Studies show that when treating cervical chordomas, en bloc resection of tumors is associated with better oncologic outcomes compared to intralesional resection.[3]

## INITIAL PRESENTATION

A 48 year old male with a history of hypertension and hyperlipidemia was admitted to Cooper Hospital for left arm pain and weakness that began in 3/2022. The patient endorsed neck and left shoulder pain that was 5/10 in severity, aching but sharp with movement, and was associated with left upper extremity (LUE) numbness and tingling. The patient also endorsed a new onset of throat tightness when swallowing food which started acutely prior to admission. The patient tried meloxicam and cyclobenzaprine when his symptoms first began without relief. On presentation, the patient had a negative surgical history and did not use tobacco or alcohol.

## PHYSICAL EXAM

A physical exam revealed left shoulder tenderness with limited range of motion (ROM) in the left shoulder. The patient's neck was supple with pain on palpation of trapezius trigger points on the left side and fullness on the left neck at level II/III, the upper jugular and middle jugular lymph nodes respectfully. There was no weakness with 5/5 strength in the upper and lower extremities, sensation was intact to light touch, reflexes were symmetrical throughout, and there was no clonus or Hoffman's sign. The patient had dysesthetic pain in the left C5 dermatome. A flexible fiberoptic nasopharyngolaryngoscopy revealed fullness of the left posterior oropharyngeal and hypopharyngeal wall from the soft palate to the hypopharynx.

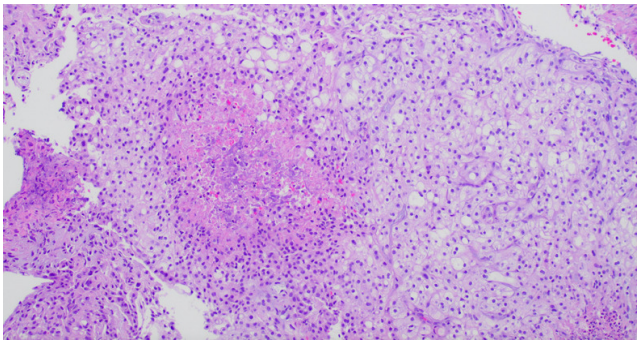

**Figure 2. Hematoxylin and eosin staining of the transoral biopsy.**

## DIAGNOSTICS

The patient's preoperative CTA neck **(Figure 1)** revealed a 5 cm left paraspinal mass with extension to the left post-styloid parapharyngeal space from C2-6 with anterior left sided erosion of the C3 vertebral body and narrowing of the left C4-C5 transverse foramina. There was encasement and narrowing of the left vertebral artery.

No metastatic disease was detected on a preoperative CT chest, abdomen, and pelvis.

A preoperative MRI cervical spine with and without contrast revealed a T1 hypointense, T2 hyperintense, heterogeneously enhancing mass. The 6.7 centimeter, necrotic mass was in the left anterior prevertebral region and extended from C2-C6, next to the left aspect of the oropharynx and hypopharynx. There was local extension into the C3-5 neural foramina and the left epidural space at C3-5. The C3-C5 vertebral bodies showed narrowing and erosion, and the C4-C5 transverse foramina showed narrowing. Encasement and narrowing of the left vertebral artery was confirmed in the C3-6 level.

A direct laryngoscopy with transoral biopsy was performed, which revealed a pale, firm, and rubbery tumor that was sent for tissue sampling (Figure 2). The specimen was positive for Brachyury, a gene that is diagnostic of Chordomas.

## TREATMENT

We aimed for gross total resection of the tumor using a combined anterior and posterior approach. Given arm weakness, the patient was prepared for the possible severity of injury to or sacrifice of the cervical rootlets to his arm. Preoperative embolization of the vertebral artery, which passed through the center of the tumor, was performed. In the operating room, a transcervical cut was made on the left side of the neck (Figure 3). The sternocleidomastoid and carotid sheath were retracted laterally, and the laryngeal framework was retracted medially. This revealed the mass underneath the prevertebral fascia. The fascia was then incised, and the tumor was dissected circumferentially. The vertebral artery was identified with embolization material within and was resected with the specimen (Figure 4). The tumor was resected from the anterior surface of the C2-C5 vertebral bodies. In order to allow for removal of the tumor, the anterior component was transected at this time. We then removed tumor from the cervical spinal bone and performed diskectomies on C3-4 and C4-5. Implants were placed into the disc spaces at both the C3-4 and C4-5 levels.

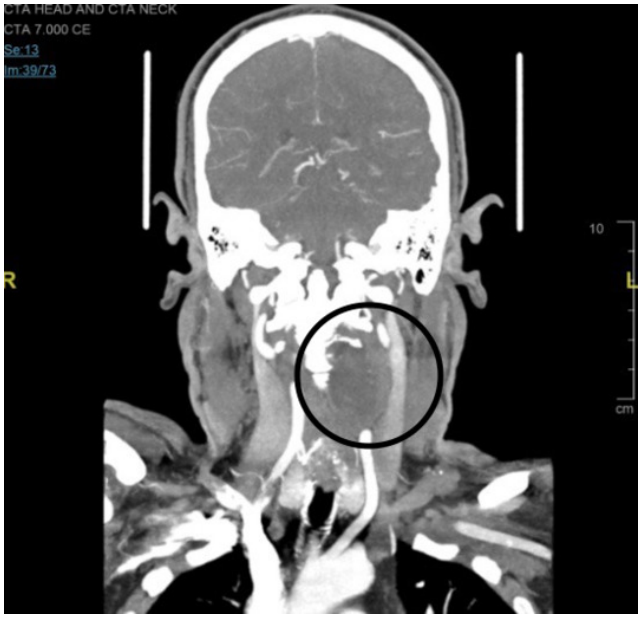


**Figure 1. CTA head and neck with a paraspinal mass shown on the left side encasing the left vertebral artery from the coronal plane.**

The patient was rearranged for a posterior approach. The C2-C6 laminae were exposed, and pilot holes were drilled into C3-C6, except C4 on the left side. Screws were then placed at C3-C6 bilaterally, except C4. The spinous processes of C3-C5 were removed and foraminotomies were performed at C3-4 and C4-5 on the left, which freed the C4 lateral mass. The epidural tumor encasing the left C4 nerve root was dissected off the dura. The C4 nerve root was resected after it was stimulated with no discernible response. All tumor was removed other than a small area in the left paraspinal region ventrolateral to the spinal column from C4-C5, which was planned to be treated with adjuvant radiation to preserve the neurovascular bundles in the spine, which would have required sacrifice in order to completely clear the tumor. Rods were attached to the screws from C2 to C6 and Medtronic Magnifuse was placed from C2 through C6. Vancomycin powder was added to the wound and a hemovac drain was placed.

## FOLLOW UP

The patient recovered from the surgery with no radiculopathy and no loss of strength in their upper extremities. A post-operative sagittal CT through the middle of the vertebral bodies showed stand-alone interbody spacer placement at C3-C4 and C4-C5, and at the levels of the laminectomy from C3-C5. Post operative right and left sagittal CT

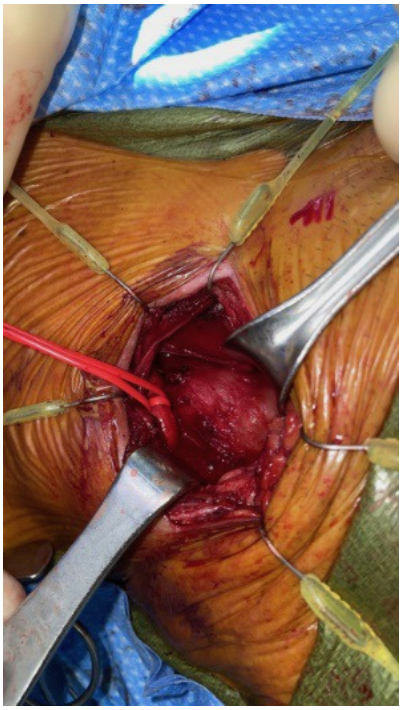

**Figure 3. Intra-operative photo from the anterior approach with visualization of the chordoma mass and left common carotid artery encircled by the red vessel loop.**

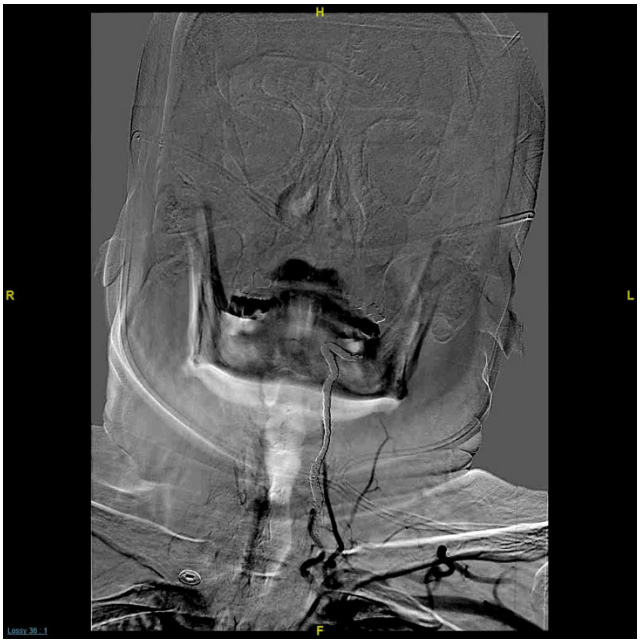


**Figure 4. Mid-arterial phase image of the left vertebral artery after his successful balloon test occlusion and subsequent coil embolization.**

scans showed hardware placement from C2-C6 and resection of the C4 lateral mass (**Figure 5**). Three month post operative AP and lateral x rays revealed adequate hardware placement and cervical alignment.

The patient presented 6 weeks post operatively and was diagnosed with sub-acute meningitis. Cultures from a lumbar puncture were positive for Streptococcus anginosus, but no growth was found on cervical spine biopsy or a thoracic fluid collection. The patient was admitted, treated with antibiotics, and discharged with a 6-week course of IV ceftriaxone. The meningitis was most likely secondary to the invasive procedure. Prompt recognition and treatment of post operative infections is necessary for patients with surgeries to remove cervical chordomas.

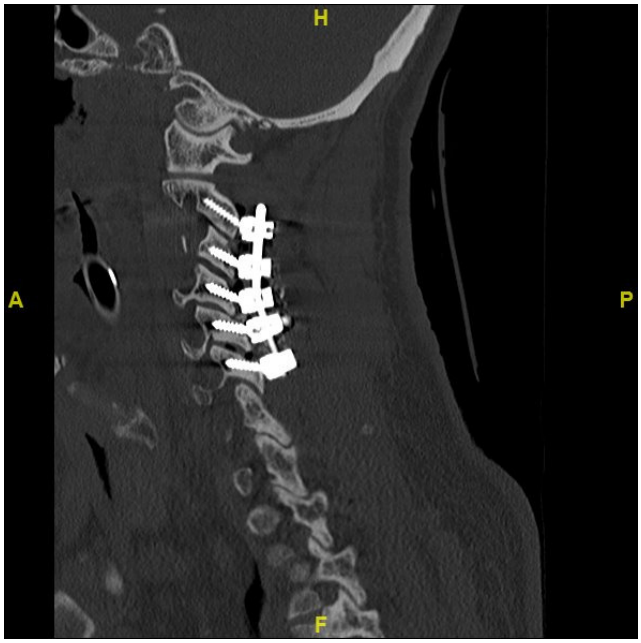


**Figure 5. Post-operative sagittal CT scan through the right sided lateral mass.**

The patient completed 6 weeks of radiation therapy, which included a course of proton beam radiotherapy. A six month post operative MRI revealed an area of stable, enhancing soft tissue in the paraspinal region of C4-C5, which is concerning for residual tumor, consistent with prior MRIs. The patient will continue following up with serial imaging to monitor for tumor recurrence and/or progression.

## DISCUSSION

Diagnosis of a chordoma is made mainly via histological staining for Brachyury, a transcription factor required for axial development and the formation of the posterior mesoderm. Brachyury expression is lost as the notochord develops and is typically undetectable ten weeks after birth.[4] Abnormal Brachyury expression is highly associated with chordomas.[4] Brachyury activates YAP (Yes-associated protein), which is a master regulator of organ development, and through this mechanism, Brachyury leads to tumor growth and epithelial mesenchymal transformation.[5] In a study that analyzed 53 chordomas, 323 non chordoma neoplasms, and 33 normal tissues, Brachyury was found to be expressed in all chordomas, labeling both chondroid and chordoid areas, and absent in all other neoplasms and non-neoplastic tissues.[6]

This case described the subtotal resection of a chordoma using a combined anterior and posterior intralesional approach. This approach involves dissecting the tumor capsule and can be classified as gross total resection or subtotal resection, depending on if all the tumor cells are removed. En bloc resection is preferred if possible and is associated with lower recurrence rates (13% of chordoma patients treated with en bloc resections had recurrence while 45% of patients with intralesional resections had recurrence[7]). However, for complicated lesions, this is often not possible and subtotal resection with adjuvant radiation for high-risk areas offers a less morbid alternative with good rates of tumor control and functional success.

76% of operations to remove cervical chordomas are performed using the intralesional technique. This is due to encasement of crucial anatomical structures by the tumor, mainly the vertebral arteries and the cervical nerve roots. Chordomas are slow growing tumors that typically do not cause symptoms until they have significantly progressed, and in many cases, have already encased the vertebral arteries or cervical nerve roots. This is a challenge for surgeons because while the en bloc technique leads to lower recurrence rates, it also results in more postoperative complications. Removing the vertebral artery predisposes patients to strokes and spinal cord ischemia, and removing nerve roots can lead to debilitating neurological deficits.[3]

In this case, we performed a subtotal resection to preserve the neurovascular bundles in the spine that would have required sacrifice to completely remove the tumor. The patient's left vertebral artery was encased by the tumor, and it was sacrificed after passing a balloon test occlusion. The left C4 nerve root was resected after it was stimulated with no discernable response.

Following adjuvant radiation therapy, imaging shows stable residual disease. The patient has normal range of motion and 5/5 muscle strength in all extremities.

## CONCLUSION

Cervical chordomas are rare, difficult to treat malignancies. It is important to keep cervical chordomas in differential diagnoses for patients with a neck mass. This case demonstrates the complex multidisciplinary surgery and necessary post operative care required to effectively treat cervical chordomas.

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