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C2 Lesion in An Adolescent Resembling Chordoma : Histophological Confirmation of Rheumatoid Pannus Following Two-Stage Decompression : A Case Report

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ABSTRACT

Introduction: Chordomas prevalence peaks in men around late middle age. Mostly occur in the sacrococcygeal and around base of the skull. Chordoma of the cervical spine is observed only in 6% of all chordoma. Growth slowly in size before becoming symptomatic and encase surrounding vascular and nerve structures.
Case Presentation: A 17-years-old male with weakness in all extremities since 2 months with history of fell to the ground with his back landed first after somersault. After that, he began to fell numbness in his all extremities. Urinary, fecal incontinence, and decrease of libido was found. Patient undergo CT scan and MRI that show canalis stenosis C2 d/t C2 Fracture Levine classification type 1A ASIA D dd spondilitis TB C2 dd susp. Pannus ec Rheumatoid arthritis. Patient underwent the occipitocervical fusion. So he underwent the second operation which was trans-oral decompression and the pathology impression was chordoma.
Discussion: Chordomas are a family of primary bone tumors, originate from undifferentiated embryonic notochord remnants present in the midline, extending throughout the skull base and axial skeleton. Only 6% of all chordoma cases mainly affect the cervical spine, as chordomas can often be found in the cervical vertebrae, either a new primary tumor or a metastatic tumor
Conclusion: Chordoma is a malignant neoplasma that if the site lesion was ini cervical, the chief complaint was the effect of spaced-occupying lesion cervical, like weakness ini all extremities
Keyword: Chordomas, Adolescent, Two-Stage Decompression

1. Introduction

Chordomas are rare malignant neoplasms with an estimated incidence of less than 0.1 per 100,000; this prevalence peaks in men around late middle age [1,2]. They account for1–4% of all primary bone tumors. Most occur in the sacrococcygeal region and around the base of the skull. Chordoma of the cervical spine is observed only in 6% of all chordomas, and in these cases, the lesion is mainly located in the midline. These tumors slowly grow in size before becoming symptomatic and encase surrounding vascular and nerve structures. Patients with advanced chordoma have a poor prognosis due to local recurrence with infiltration and destruction of adjacent bone and tissues. As systemic chemotherapy options have been found to be not fully effective, molecular targeted therapies, especially tyrosine kinase inhibitors, have been proposed [3]. However, despite significant advances in therapeutic options, radical resection with subsequent postoperative radiotherapy remains the mainstay of care [1,4–6].

2. Case Presentation

A 17-year-old male presented with complaints of progressive weakness in all four extremities that had started one and a half months prior to admission. The weakness was insidious in onset, beginning in the lower limbs and ascending to involve the upper limbs. He reported difficulty in ambulation, frequent falls, and inability to hold objects. Two months prior to admission, the patient began to experience numbness in all extremities. Over time, he also developed bladder and bowel dysfunction, characterized by difficulty voiding, use of catheter, and constipation. Additionally, the patient reported erectile dysfunction and loss of perianal sensation.

Three months before admission, the patient experienced neck pain following a sports-related injury during a somersault, where he landed on his back with significant force. He did not seek immediate medical evaluation and instead underwent traditional massage therapy. Post-injury, he complained of stiffness and reduced range of motion in the cervical region.

There was no history of fever, night sweats, weight loss, chronic cough, or known contact with tuberculosis. No history of tuberculosis treatment or underlying pulmonary or systemic disease was noted.

Neurological examination showed tetraparesis with motor strength graded 2/5 across all myotomes in both upper and lower extremities. Hypoesthesia was noted below the C3 dermatome. Deep tendon reflexes were hyperactive (+++/+++) with positive Babinski and clonus bilaterally. Physiological reflexes were exaggerated. The bulbocavernosus reflex was present. The ASIA classification was D.

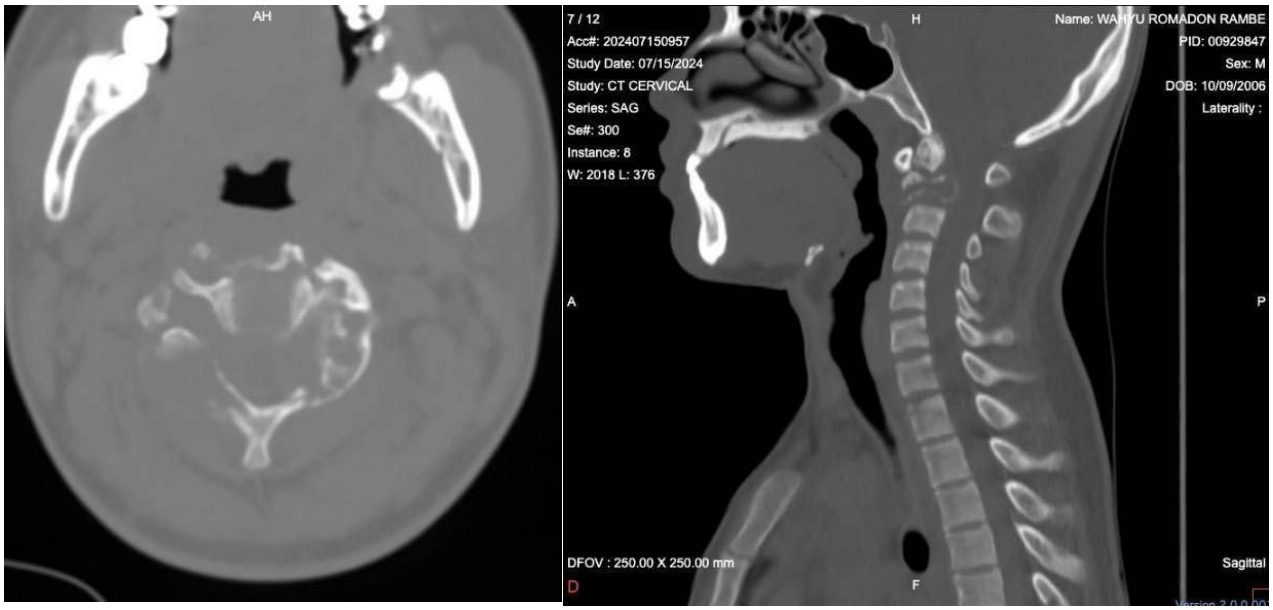

Figure 1. Axial CT image of the C2 vertebra demonstrating a burst fracture involving the vertebral body, lamina, and pars interarticularis. The comminution and canal compromise are evident, consistent with Levine Type 1A classification. No atlantoaxial dislocation is seen.

Imaging studies were performed. MRI of the cervical spine on July 1, 2024, showed canal stenosis at the C2 level, a compression fracture of C2 vertebra, and posterior disc protrusion causing anterior spinal cord compression. CT cervical spine performed on July 15, 2024, confirmed a fracture involving the corpus, pars interarticularis, and lamina of the C2 vertebra, consistent with a Levine Type 1A injury. There was no atlantoaxial dislocation.

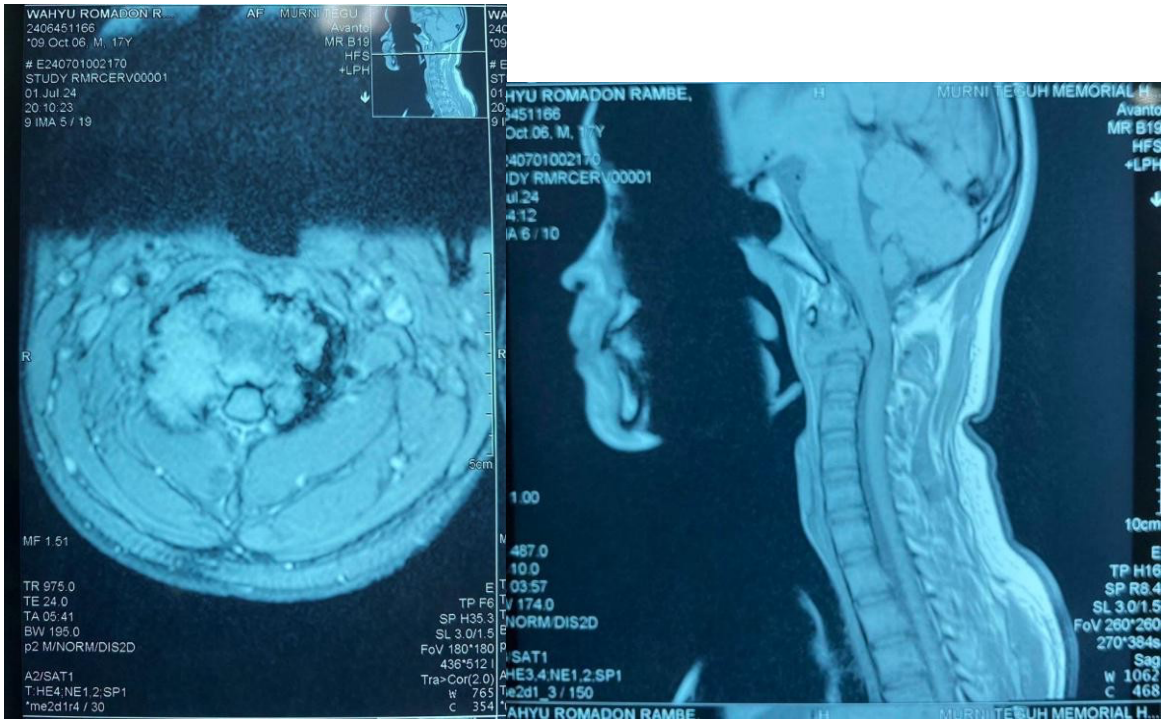

Figure 2. T2-weighted sagittal MRI showing canal stenosis at the C2 level with a burst fracture of the C2 vertebral body. Note the anterior spinal cord compression and hyperintensity within the cord, suggestive of spinal cord edema or early myelomalacia. The posterior disc protrusion is seen impinging on the dural sac.

Based on clinical and radiological findings, the diagnosis of cervical canal stenosis due to C2 fracture with resulting tetraparesis (ASIA D) was made. Differential diagnoses included tuberculous spondylitis and pannus formation due to rheumatoid arthritis, although these were considered less likely due to lack of supporting systemic signs and laboratory parameters.

The patient underwent urgent occipitocervical fusion from the occiput to C3 on September 5, 2024. Intraoperatively, instability at the C2 level was confirmed, and instrumentation was applied to stabilize the craniocervical junction. Postoperatively, the patient received ceftriaxone 1 g every 12 hours, ketorolac 30 mg every 8 hours, ranitidine 50 mg every 12 hours, and intravenous fluids. He was immobilized using a rigid cervical collar. Pain was assessed using the visual analogue scale (VAS) and rated as 2–3. Early rehabilitation and passive limb movements were initiated.

Histopathological examination of the tissue from the C2 vertebral body revealed fibrous and granulomatous tissue with epithelioid cells and histiocytes but no evidence of malignancy. The lesion was interpreted as pannus consistent with inflammatory or autoimmune etiology. However, the granulomatous architecture and radiological pattern necessitated exclusion of atypical entities such as chordoma, which is known for causing lytic bone destruction and mimicking granulomatous masses.

The patient was discharged in stable condition with ongoing physiotherapy and neurorehabilitation. Two months later, on November 2, 2024, the patient returned to the emergency department with complaints of intermittent shortness of breath for the past week. He also reported no significant improvement in limb weakness and persistent bladder and bowel dysfunction. On examination, breath sounds were decreased bilaterally, with no added sounds. Oxygen saturation was 99% on nasal cannula at 3 L/min. Neurological status remained ASIA D with 1/5 to 2/5 motor strength, persistent hypoesthesia below C3, and positive Babinski sign. No new deficits were identified. A chest X-ray showed no acute pulmonary process. The patient was readmitted for evaluation and continued supportive therapy.

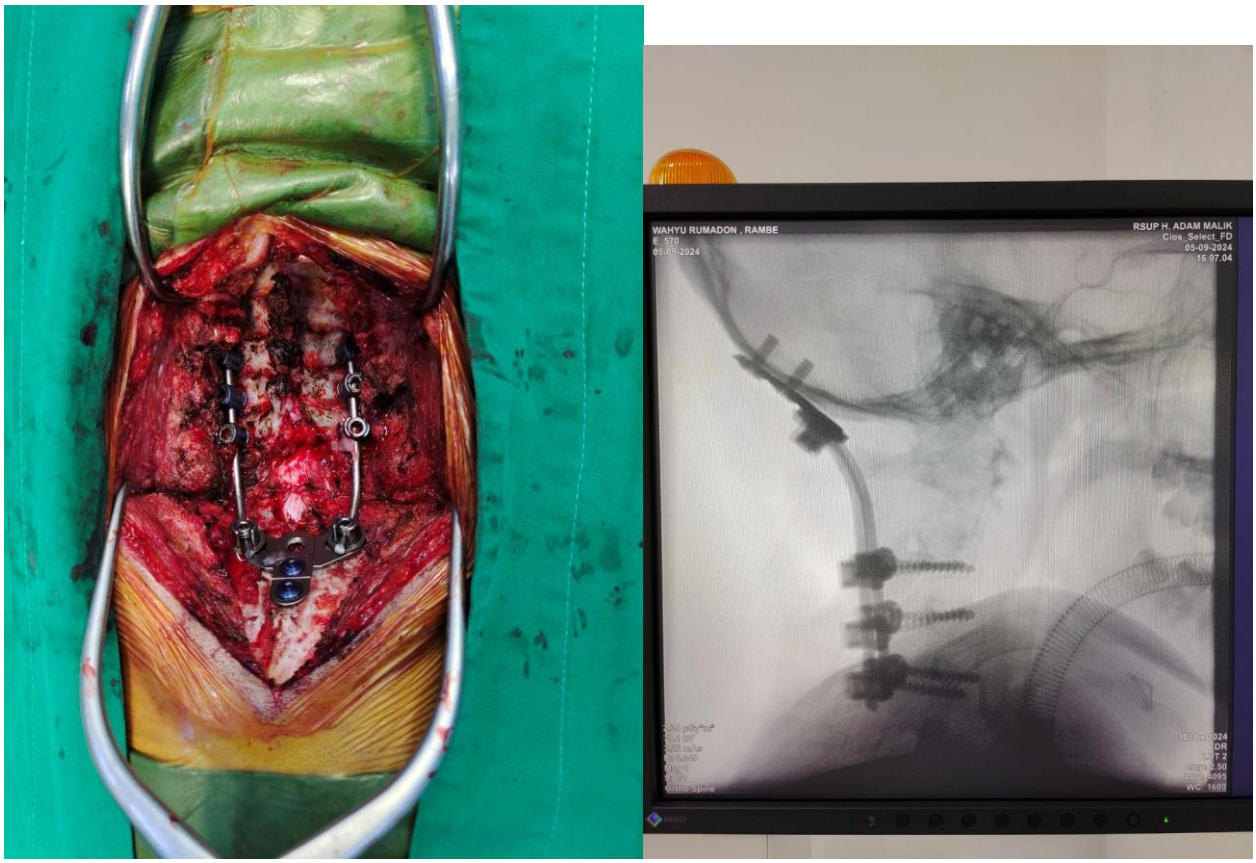

Figure 3. Lateral plain radiograph of the cervical spine post occipitocervical fusion, showing proper placement of occipital and cervical instrumentation from the occiput to C3. The alignment is restored, and the hardware maintains stability across the craniocervical junction.

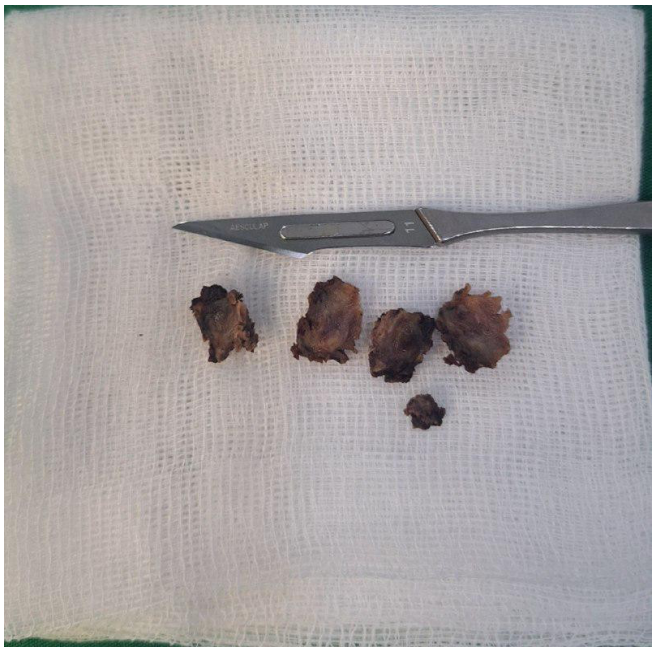

Figure 4. Histopathological section of the C2 lesion showing fibrous granulation tissue with fibrin deposition and clusters of epithelioid histiocytes (H&E stain, 400×). No evidence of caseating necrosis or malignant cells. Findings are consistent with pannus formation due to rheumatoid arthritis.

3. Discussion

Upper cervical spine lesions involving the C2 vertebra are uncommon in adolescents and can represent a broad range of pathologies, including traumatic, inflammatory, infectious, and neoplastic etiologies. This case initially presented as cervical myelopathy secondary to a Levine type IA C2 fracture, but the accompanying soft tissue mass and persistent neurologic deficits prompted consideration of other differential diagnoses such as tuberculous spondylitis, rheumatoid pannus, and neoplasm—particularly chordoma, a rare yet critical entity to exclude in this context.

Chordoma is a malignant bone tumor derived from embryonic notochordal remnants, comprising less than 5% of all primary bone tumors. Although most commonly found in the sacrum (50%) and clivus (30%), approximately 10–15% occur in the mobile spine, with the C2 vertebra being a recognized site of involvement [7,8]. Chordomas typically affect adults between 40 and 60 years old, but cases in children and adolescents have been reported, albeit rarely [9] Given its location and indolent progression, chordoma of the upper cervical spine often mimics other pathologies, including rheumatoid pannus, tuberculosis, and chronic post-traumatic changes.

Clinically, chordomas at the C2 level often present with insidious neck pain, progressive myelopathy, and sometimes lower cranial nerve involvement. In our case, the patient exhibited progressive quadriparesis, sensory deficits, and autonomic dysfunction. Radiologically, chordomas usually appear as lytic, expansile lesions with bony destruction and soft tissue extension, often centered in the vertebral body. MRI typically demonstrates a lobulated mass that is hypointense on T1-weighted and hyperintense on T2-weighted sequences, with moderate to marked post-contrast enhancement [9,10]. These features, while suggestive, are not specific and frequently overlap with those of inflammatory lesions, particularly in the context of trauma or suspected infection.

In this case, although the imaging showed canal stenosis with vertebral collapse and a soft tissue component, the absence of aggressive bone lysis and paraspinal extension made chordoma less likely. However, due to the patient's neurological deterioration despite posterior stabilization, a second-stage anterior decompression was indicated. This approach is also consistent with the standard of care in resectable chordomas of the upper cervical spine, which often require a multidisciplinary strategy including gross total resection and adjuvant radiotherapy with proton or carbon ion therapy [8,11].

Histopathologic confirmation remains the gold standard for diagnosing chordoma. Microscopically, chordomas are characterized by physaliphorous cells—vacuolated tumor cells arranged in cords within a myxoid matrix—and demonstrate immunopositivity for brachyury, cytokeratin, EMA, and S100 protein [10, 12]. In our patient, histological examination showed granulomatous inflammation with fibrin and epithelioid histiocytes, without signs of malignancy or notochordal differentiation, thus excluding chordoma.

Although rare, the inclusion of chordoma in the differential diagnosis is essential, especially in adolescents presenting with upper cervical spinal cord compression and mass lesions. The distinction is critical because the prognosis, surgical approach, and adjuvant therapies differ significantly from those for inflammatory or infectious lesions. Early diagnosis and radical resection offer the best outcomes, as chordomas are notoriously resistant to chemotherapy and conventional radiation [8,11].

This case underscores the importance of maintaining a high index of suspicion for chordoma in upper cervical lesions and highlights the necessity of integrating clinical, radiologic, and histopathologic data to achieve a definitive diagnosis. In patients with incomplete neurologic recovery after posterior fusion and persistent anterior compression, timely anterior decompression and biopsy not only serve therapeutic goals but also play a diagnostic role in differentiating chordoma from its mimics.

4. Conclusion

This case illustrates the importance of considering a broad differential diagnosis—including trauma, infection, inflammation, and neoplasm such as chordoma—in adolescents presenting with cervical myelopathy and C2 lesions. Although initial imaging suggested a stable fracture, persistent neurological deficits and anterior compression required further intervention. A staged surgical approach with posterior fusion followed by transoral decompression proved essential for both treatment and diagnosis. Histopathology confirmed pannus formation due to rheumatoid arthritis, highlighting the need for tissue confirmation in ambiguous cases. Early recognition and appropriate surgical management are critical to prevent permanent spinal cord injury.

Acknowledgements

None.

Conflict of Interest

The authors declare no conflicts of interest in preparing this article.

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