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Cervical Spine Malignancy in Adults: Challenges with Multidisciplinary Approach - A Case Report

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Abstract

The upper cervical spine is a rare but critical site for pathological fractures secondary to neoplastic processes. Lesions involving C2 and adjacent vertebrae pose significant risks of instability, cord compression, and neurological deterioration, requiring prompt surgical intervention. We report a 47-year-old male who presented with a 2-month history of neck pain and headaches, followed by progressive weakness in both upper and lower limbs and progressive voice loss over 1 month. Imaging revealed an expansile lytic lesion of the C2 vertebra involving the odontoid process, associated with a pathological fracture. Additional findings included a lytic lesion in the superior body of C3 and a large prevertebral soft tissue mass measuring 6.8 × 3.4 cm. The atlanto-occipital joint was preserved. Given the instability and progressive neurological compromise, the patient underwent posterior decompressive laminectomy at C1 and C2. Instrumented stabilization was achieved using C2 pars screw on the left, bilateral lateral mass screws on C3 and C4, and exclusion of C1 and right-sided C2 fixation due to anatomical constraints. The procedure achieved adequate decompression and stabilization of the cervical spine. Postoperative recovery was uneventful, with early neurological improvement. Histopathological evaluation of the lesion reviewed an adenocarcinoma initially but later was confirmed to be a chordoma. This case highlights the importance of timely recognition and surgical stabilization of destructive cervical spine lesions. Decompressive laminectomy combined with lateral mass fixation provides effective neural decompression and spinal stability in complex upper cervical

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Introduction

Tumours of the upper cervical spine are uncommon but present disproportionate clinical and surgical challenges due to the region's complex anatomy, proximity to the brainstem, cranial nerves, and vertebral arteries, and the functional importance of the atlanto-axial complex. Chordomas are malignant neoplasms arising from residual notochordal tissue and are rare along the craniospinal axis and even more uncommon in the cervical region, where they classically produce expansile, lytic bone destruction with frequent soft-tissue extension and a high risk of local recurrence if not managed aggressively. Because chordomas are relatively chemo-resistant, maximal safe surgical resection combined with adjuvant radiation (often proton or heavy-ion therapy where available) remains the cornerstone of durable local control [1,2].

Chordoma, by contrast, is a rare primary malignant tumor of notochordal origin that most often arises in the sacrum and skull base, with the mobile spine (including the cervical segment) less frequently affected. Despite its rarity, cervical chordoma carries substantial morbidity due to local aggressiveness and a propensity for recurrence; recent multicenter data underscore the guarded prognosis and highlight the need for disease-specific management [2]. Diagnostic confusion can occur because chordomas often express epithelial markers (e.g., cytokeratin, EMA) and may be initially interpreted as metastatic adenocarcinoma on small biopsies particularly when the lesion is midline and lytic. Brachyury (TBXT) nuclear immunoreactivity is highly sensitive and specific for chordoma and is therefore pivotal in resolving this differential. Case reports

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document misclassification of chordoma as adenocarcinoma, reinforcing the importance of a targeted immunohistochemical panel that includes brachyury when evaluating cervical destructive lesions with epithelial marker positivity [3,4]

The distinction is not academic: treatment paradigms diverge substantially. While metastatic adenocarcinoma to the cervical spine is commonly managed with systemic therapy, radiotherapy, and selective stabilization, optimal chordoma care prioritizes maximal oncologic resection when feasible and high-dose conformal radiotherapy (proton or carbon-ion) for local control; extent of resection correlates with survival and local recurrence. Accurate early classification therefore directs surgical planning and referral for particle therapy, ultimately influencing function and outcomes [5-7]. This case, which was initially labeled as adenocarcinoma but ultimately proven to be chordoma, illustrates a high-stakes diagnostic pitfall in cervical oncology. Embedding brachyury testing and chordoma-aware radiologic, pathologic correlation into the workup of midline C2-C3 lytic masses can prevent misdiagnosis and ensure timely, disease-appropriate management [3].

Beyond diagnosis, the cervical location (especially C1–C3) raises important multidisciplinary surgical questions. Tumours centered at the craniovertebral junction or high cervical spine often cross the traditional domains of skull-base, headand-neck, and spine surgery: anterior exposures (transoral, endonasal, or lateral approaches) may be most familiar to head and neck / otolaryngology and skull-base teams, while posterior decompression and instrumented stabilization are typically in the neurosurgical and spinal orthopaedic domain. Complex cases therefore demand coordinated planning among otolaryngologists/head and neck surgeons, neurosurgeons (or spine surgeons), radiologists, radiation oncologists, and neuropathologists to achieve both oncologic control and mechanical stability while minimizing neurological morbidity. Contemporary series underline that outcomes, extent of resection, neurological recovery, and recurrence rates are best when care is delivered by experienced, multidisciplinary teams with access to high-precision radiotherapy and formal preoperative planning conferences [8,9].

Case

We present the case of a 47-year-old male from Victoria Falls with no significant past medical or surgical history apart from well-controlled hypertension. He was previously well until he developed progressive neck pain and headaches over a twomonth period. One month later, he experienced worsening weakness in both upper and lower limbs, progressing to the point where he could no longer walk or sit unaided. During the same period, he noted progressive deterioration of his voice, which declined to a whisper. On examination, the patient was alert with a Glasgow Coma Scale score of 15/15 and pupils equal and reactive to light. Neurological assessment revealed symmetrical weakness in both upper and lower limbs, graded at 2/5 on the Medical Research Council (MRC) scale. Sensory function was preserved, and bladder and bowel control remained intact. Cranial nerve examination showed loss of vocal pitch, suggestive of laryngeal nerve involvement. Systemic examination was otherwise unremarkable. This

constellation of progressive neck pain, neurological decline, and voice changes in an otherwise healthy middle-aged man prompted urgent imaging, which revealed destructive lytic lesions in the upper cervical spine with associated pathological fracture and prevertebral soft tissue mass, consistent with an underlying malignant process.

Surgical Procedure

The patient was positioned prone under general anesthesia, and the posterior cervical region was prepared and draped in the usual sterile fashion. A midline posterior incision was made, extending from the inion to the spinous process of C4. Subperiosteal dissection exposed the posterior elements of the craniovertebral junction and upper cervical spine. A decompressive laminectomy of C1 and C2 was performed to achieve adequate neural decompression. Posterior stabilization was then undertaken using lateral mass and pars fixation. A left-sided C2 pars screw was successfully inserted, while screw placement was not feasible on the right C2 or at C1 due to anatomical constraints. Bilateral lateral mass screws were placed at C3 and C4, providing stable fixation. Adequate decompression and stabilization of the cervical spine were confirmed intraoperatively. Hemostasis was achieved, the wound was closed in layers, and the patient was transferred to recovery in a stable condition.

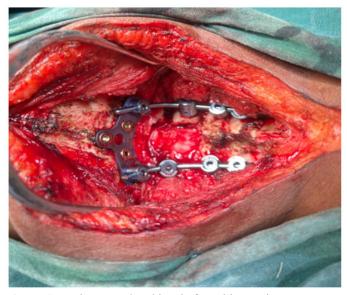


Figure 1: Fusion completed just before skin repair



Figure 2: Post operation imaging with instrumentation in-situ

Progress

The patient was monitored in the intensive care unit for 3 days following surgery and subsequently transferred to the general neurosurgical ward in stable condition. His postoperative course was uneventful, and he demonstrated gradual neurological improvement. By the time of discharge on postoperative day 14, muscle power in both upper and lower limbs had improved from a preoperative grade of 2/5 to 3/5 on the Medical Research council (MRC) scale. Notably, there was marked improvement in vocal strength, with restoration from whispering to a nearnormal voice pitch. At follow-up two weeks after discharge, the patient continued to show neurological recovery with stable wound healing, improved limb strength, and sustained improvement in phonation. No new deficits or complications were observed.

Discussion

The case, showed an expansile, lytic midline lesion of C2 involving the odontoid with a large prevertebral soft-tissue component, initially reported as adenocarcinoma but later identified as chordoma on biopsy taken at fusion, it illustrates several important diagnostic and management principles for lesions of the upper cervical spine. The initial management was undertaken by otorhinolaryngology (ENT) surgeons due to predominant airway and prevertebral mass concerns. However, given rapid neurological deterioration and radiological evidence of cord compromise, care was later assumed by the neurosurgical team.

Firstly, the clinical–radiologic pattern strongly favours chordoma. Chordomas are rare, notochord-derived neoplasms that classically arise in midline axial sites (clivus, mobile spine, sacrum) and often present as slow-growing, locally destructive, expansile lesions with bone lysis and soft-tissue extension. Cervical chordomas, while uncommon relative to sacral and clival sites, are well described and are notorious for causing local instability and neurological compromise when they involve C2/odontoid structures. These anatomic and radiologic features which are; midline location, involvement of the odontoid, expansile lytic destruction with a sizeable prevertebral soft-tissue mass, are all typical of chordoma rather than most metastatic adenocarcinomas, which more commonly produce multifocal metastatic deposits and a different clinical course [2,10].

Secondly, histopathology and immunophenotype explain the initial diagnostic confusion. Chordoma cells frequently coexpress epithelial markers (pan-cytokeratin, EMA) and S100/vimentin, which can mimic metastatic adenocarcinoma on small or limited biopsy specimens and on immunostains that are restricted to epithelial markers. This overlap is a well-recognized pitfall: small samples taken before a full immunohistochemical panel is applied may be labelled "adenocarcinoma" when only epithelial markers are tested. The discovery of brachyury (TBXT) nuclear expression revolutionized chordoma diagnostics, brachyury is highly sensitive and specific for notochordal-derived tumors and distinguishes chordoma from metastatic epithelial malignancies that lack nuclear brachyury. In practice, a midline axial lytic lesion that shows cytokeratin/EMA positivity should prompt reflex testing for brachyury to

avoid misclassification. The positive brachyury result on the intraoperative/fusion biopsy in our patient therefore provided the clinico-pathologic concordance that was lacking in the original report [11,12].

The diagnostic distinction matters because it changes oncologic strategy. Management paradigms diverge: metastatic adenocarcinoma to the spine is generally treated with systemic therapy, palliative radiotherapy, and stabilization when indicated, whereas chordoma is best managed, when anatomically and physiologically feasible, by maximal oncologic resection (ideally en-bloc with wide margins in sacral/clival sites) together with high-dose conformal particle radiation (proton or carbon-ion) to achieve durable local control. Mobile-spine chordomas are frequently not amenable to true en-bloc excision because of epidural invasion and the proximity of neurovascular structures; consequently, intralesional or staged decompression/stabilization procedures are often performed as "separation" or palliative surgeries, with adjuvant high-precision radiotherapy providing the best chance of local disease control. Our patient's posterior decompressive laminectomy and lateral-mass fixation reflect such a balance between urgent neurologic decompression and the anatomic limits to radical excision in the upper cervical spine [5-7].

Fourth, contemporary adjuncts and planning considerations follow from these realities. For mobile-spine chordoma patients who undergo instrumented stabilization, implant selection and radiotherapy planning are important: carbon-fiber-reinforced PEEK implants have been proposed to improve radiotherapy dosimetry and postoperative imaging surveillance compared with standard titanium constructs. Moreover, because chordomas are relatively insensitive to conventional photon radiotherapy, referral for particle therapy (proton or carbon ion) should be considered early, especially when en-bloc resection is not achievable. Multidisciplinary discussion, neurosurgery/spine surgery, radiation oncology experienced in particle therapy, medical oncology, and pathology, optimizes both local control and functional outcome [7,13].

Finally, this case highlights practical lessons for clinicians in resource-constrained settings. First, maintain a high index of suspicion for chordoma in midline axial lesions with odontoid involvement and expansive soft-tissue components, even when early biopsy suggests carcinoma. Second, ensure that initial histopathology includes or reflexively progresses to a chordoma panel (brachyury ± cytokeratin, EMA, S100) whenever the radiographic pattern is concordant, this reduces misdiagnosis and avoids inappropriate systemic-therapy referrals. Third, recognize that urgent posterior decompression and stabilization, as performed here, can be life- and functionsaving when instability or cord compression is present; subsequent oncologic care (definitive resection or particle radiotherapy) should follow once diagnosis is secured [11,12].

Conclusion

This case highlights the diagnostic challenges of destructive cervical spine lesions, particularly when initial histology suggested adenocarcinoma. The subsequent biopsy obtained intraoperatively confirmed a chordoma, a diagnosis more consistent with the clinical and radiological picture of a midline expansile lytic lesion centered on the C2 vertebra with associated prevertebral soft tissue mass. Unlike metastatic adenocarcinomas, chordomas characteristically arise in the axial skeleton, demonstrate slow but locally aggressive growth, and frequently present with pathological fractures and progressive neurological compromise. Immunohistochemical confirmation with brachyury further distinguishes chordomas from adenocarcinoma. Accurate differentiation is critical, as management strategies and prognostic implications diverge significantly: adenocarcinomas often indicate systemic disease requiring palliative approaches, whereas chordomas demand maximal surgical resection and high-dose conformal radiotherapy for local control. This case underscores the importance of correlating histological findings with clinical and radiological features to avoid misclassification and to ensure appropriate, disease-specific treatment planning.

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