

Technical Notes & Surgical Techniques

Axial chordoma of the cervical spine managed by a trans-mandibular trans-oral approach

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A B S T R A C T

Chordomas are low-grade but locally aggressive malignant neoplasms derived from notochordal remnants. These lesions are best treated by en bloc however this is especially challenging in the upper cervical spine due to the proximity of vital structures.

We present a 46 year old male whom presented to our unit complaining of progressive upper cervical axial neck pain aggravated by neck rotation, of 4 months duration. He had no symptoms of myelopathy. Examination revealed high posterior spinal tenderness and resistance to neck rotation however the patient's motor and sensory examinations were normal and he had normal gait. An inter-disciplinary team comprising Orthopedics, Neurosurgery, Maxillofacial surgery and Otorhinolaryngology took the patient for surgery. The first stage of the procedure comprised Otorhinolaryngology performing a tracheostomy and thereafter a combined Orthopedic Neurosurgical procedure involving a posterior instrumented C1–C3/C4 fusion and biopsy of the lesion. The second stage involved the maxillofacial surgeons dividing the patient's mandible to increase operative exposure. Thereafter the Orthopedic and Neurosurgical surgeons proceeded to perform a transmandibular transoral en bloc resection of the body and odontoid process of C2 which was replaced by a cage construct. The vertebral arteries were preserved and no dural breach occurred. Post operatively the patient showed no new neurology and was ambulant. He was maintained in a Philadelphia collar for 6 weeks until his fusion was complete.

In conclusion this difficult case was managed successfully by thorough pre-operative planning and inter-departmental co-operation. Fortunately the lesion was isolated to the body and odontoid process of C2 which was in the patients favor lending the case towards a favorable outcome.

1. Introduction

Chordomas are low-grade but locally aggressive malignant neoplasms derived from notochordal remnants. These lesions are best treated by en bloc resection to prevent recurrence but a difficult location in the upper cervical spine often makes this surgery challenging. Adjuvant therapies include stereotactic radiotherapy and proton beam radiotherapy and of late chemotherapy in the form of immunotherapy with Imatinib has showed promise. Unfortunately these adjuvant therapies often have limited success and are best regarded as palliative. For this reason en bloc resection with clear borders is the first line and most effective therapy and affords a patient the best chance of cure. The ability to achieve this is however strongly dependent on the size and location of the lesion. En bloc resection is especially challenging in the upper cervical spine due to the proximity of vital structures (Figs. 1–3).

2. Case description

We present a 46 year old male whom presented to our unit complaining of axial upper cervical pain for 4 months duration that was

progressive, aggravated by neck rotation, and poorly relieved by analgesia. He had no symptoms of myelopathy. Examination revealed high posterior spinal tenderness and resistance to neck rotation however the patient's motor and sensory examinations were normal (Figs. 4–6).

An interdisciplinary team comprising Orthopedics, Neurosurgery, Maxillofacial surgery and Otorhinolaryngology took the patient for surgery for a first stage posterior cervical fusion and biopsy, second stage anterior en bloc resection of this lesion, which on MRI scan was fortunately isolated to the body and odontoid process of C2 (Figs. 7–9).

The first stage of the procedure involved the Otorhinolaryngology department whom performed a tracheostomy on the patient. The second stage involved a combined Orthopedic-spine Neurosurgical-spine procedure for which the patient's head was placed in a Mayfield clamp and he was turned prone while his neck kept in a neutral position. A midline posterior neck incision was performed for a posterior instrumented C1–C4 fusion involving C1 lateral mass screws above, and C3 and C4 lateral mass screws below, which were placed using fluoroscopic imaging. A C2/C3 laminectomy was then performed and the C2 facet joints were identified. Both C2 nerve roots were identified

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Fig. 1. Pre-operative lateral C-spine X ray. Suspicious abnormality visible at atlantoaxial joint making the joint look grossly unstable.



Fig. 2. Pre-operative A/P Cervical spine X ray. Osteolytic lesion visible at atlantoaxial joint.



Fig. 3. Pre-operative A/P open mouth view. Osteolytic lesion clearly visible in central C2.



Fig. 4. Pre-operative lateral CT neck. Osteolysis of C2 body and odontoid process visible.

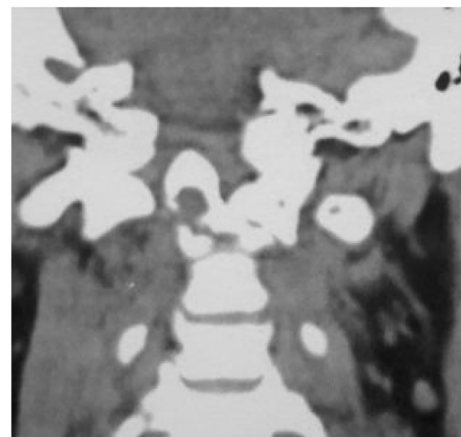


Fig. 5. Pre-operative coronal CT neck. Osteolysis of C2 body and odontoid process visible.

and transected. The vertebral arteries were identified at their entrance into the foramen transversarium of the atlas and just above the C3 superior articular process. The surgeons were now able to maintain vascular control in the event of bleeding while multiple tumor arterial feeders originating from the vertebral arteries were coagulated and

divided. The remaining accessible bone of C2 was removed on either side and a biopsy of the tumor was taken. No tumor was removed besides this small biopsy for fear of seeding the lesion posteriorly. The rods were placed and fixed and the wound was closed in layers (Figs. 10–12).

One week later the biopsy result became available confirming the lesion to be a chordoma and the patient was booked for a trans mandibular trans oral en bloc resection procedure. The patient was placed supine with his head maintained in a neutral position. The Maxillofacial surgeons now performed the third part of a staged procedure and proceeded to divide the patient's lower lip and mandible to increase operative exposure. The uvula was stitched to a Foley's catheter and pulled through the nose everting the soft palate to provide adequate visualization of the upper posterior pharyngeal wall.

Thereafter the fourth and final stage involved once more the Orthopedic and Neurosurgical surgeons whom proceeded to insert a self-retaining Crockard retractor system to maintain adequate

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