Pediatric Upper Cervical Spine Giant Cell Tumor: Case Report

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Introduction

Giant cell tumor of the bone (GCTB) is a benign bony tumor accounting for 5 to 8% of all bone tumors; it is generally observed at the epiphysis of long bones. It is rare before maturity, and it has a female predilection.1 The axial skeleton above the sacrum, especially the upper cervical spine, is an uncommon location for GCTB.1–5

Although it is benign, its locally aggressive nature makes its management demanding, especially in the upper cervical spine due to the critical surrounding neural and vascular structures. Complete en bloc excision is usually the ideal treatment option, but this method is also challenging due to its critical location.3,6 In cases with a residual tumor, radiation therapy or chemotherapy may be recommended.7

The few case reports and small series in the literature highlight the rarity of this tumor. We present this rare case involving the upper cervical spine in a 13-year-old girl that was managed surgically with a combined posterior and anterior cervical retropharyngeal approach; the tumor was determined to be successfully controlled after 2 years of follow-up.
Case Report

A 13-year-old girl, who was previously diagnosed with thoracolumbar idiopathic scoliosis, was admitted to the regional hospital. During her preoperative evaluation, she was found to have a lytic lesion in her second cervical vertebra (C2). After a full workup that included an X-ray, a computed tomography (CT) scan, a magnetic resonance imaging (MRI) scan, and a bone scan, an open biopsy of the lesion was performed using a posterior cervical laminectomy approach. A bone graft was applied, and the patient was immobilized in a halo vest. A week later, the pathology report revealed a GCTB, and she was referred to our institution for further management. A detailed history as well as general and neurologic examinations were obtained. She was neurologically intact with mild neck pain.

The cervical spine plain X-ray showed an expansile lytic lesion at C2. The CT scan revealed the details of the bony expansile lytic lesion of C2 involving the body, pedicle, and odontoid process with extensive thinning of the cortex. The lesion was associated with a soft tissue mass extending to the inside of the neuronal canal and causing effacement of the spinal cord. There was also mild C1–C2 rotational instability, as shown in Fig. 1A, B, C, and D.

The MRI with contrast revealed the extension of the lesion inside the canal with moderate compromise of the cervical spinal cord with effacement and extension of the lesion to the pedicles and lateral masses encasing both vertebral arteries. The lesion showed a low-intensity signal on T1, intermediate heterogeneous signal on T2, and intense homogenous contrast enhancement, as shown in Fig. 2A, B, C, D, and E.

A vertebral artery angiography was performed to evaluate the patency of the vessels. Both vertebral arteries were patent, but the one on the left was wider. An interventional radiologist performed a vertebral artery occlusion test, and the results revealed that both arteries were patent with good collaterals.

A musculoskeletal metastatic workup was performed; the bone scan revealed increased uptake in the C2 body and no other uptake throughout the skeleton.

The biopsy of the referring hospital was reviewed and the diagnosis was confirmed by our pathology department. However, because of the uncommon location of the lesion, the tumor board recommended another biopsy of the lesion. The interventional radiologist deferred the biopsy because of the critical location of the tumor, and he performed preoperative vertebral artery stenting to enable easy identification of the vessels during surgery and to prevent occlusion by the tumor.

An open biopsy using a posterior cervical approach with occipitocervical fusion was preferred because the spine was posteriorly explored in the initial procedure, which was done at the referring hospital. In addition, because malignancy was expected, the surgical team did not want to contaminate the anterior structures with potentially malignant cells and preferred to perform a long occipitocervical fixation down

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**Fig. 1** Computed tomography (CT) scan of the C2 axis showing the destructive lesion involving the odontoid, C2 body, and lateral masses with an element of atlantoaxial rotational instability. (A) CT scan sagittal cut; (B) CT scan coronal; (C) CT scan axial cut at the odontoid level; (D) CT scan axial cut at the C2 body level.
to C5 to achieve solid fusion, as there was no plan for another surgery at this stage.

The histopathology report confirmed the diagnosis of GCTB. The gross appearance was a firm brownish mass with foci of hemorrhaging or necrosis. Microscopically, the lesion was full of multinucleated giant cells in the mononuclear stroma, consistent with a GCTB.

After confirmation of the diagnosis of GCTB, the management plan involved complete excision of the tumor and reconstruction and stabilization of the spine, so a left retropharyngeal approach (submandibular) was preferred over a transoral approach because a wide exposure was required for the tumor excision and reconstruction using bone graft with instrumentation.

Surgery revealed a near completely destroyed C2 body and odontoid, and the tumor mass was an eggshell cortex overlapping the C3. The tumor was firm, brownish yellow, and it was friable and bled easily. Nearly complete excision of the tumor was accomplished, with excision of the C2 body, odontoid, and part of the lateral mass. Reconstruction of

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**Fig. 2** Magnetic resonance imaging of the upper cervical spine showing low signal intensity on T1, intermediate signal on T2, and intensely enhanced postcontrast. Axial cuts demonstrate an extensive soft tissue mass. (A, B) T2-weighted images, sagittal and axial cuts; (C, D) T1-weighted images, sagittal and axial cuts; (E) T1-weighted image, postcontrast, sagittal cut.

**Fig. 3** (A) Postoperative X-ray; (B) postoperative sagittal magnetic resonance imaging; (C) postoperative sagittal computed tomography scan.
the spine was performed with a fibular graft. A 3-mm-deep trough was created at the upper end of the graft to fit the anterior arch of C1 to maximize stability, and another trough was created at the body of C3 to fit the lower end of the graft. The graft was inserted by a press fit. Additional stability was applied by adding a plate from the C1 arch to the body of C3 (Fig. 3A, B, and C).

Postoperatively, the patient was transferred while intubated to the intensive care unit. The patient was extubated 2 days later. An elective tracheostomy was performed and maintained for 2 months, when it was weaned gradually until it was removed. The patient had swallowing difficulty for 3 weeks, which resolved without intervention.

Results

The final pathology supported the diagnosis of GCTB at our pathology laboratory and was confirmed by an international pathology laboratory, as shown in Fig. 4A and B. Our institution’s tumor board advised regular radiologic follow-up with no additional adjuvant therapy.

The patient was regularly followed every 6 months with no evidence of recurrence or cervical spine instability for 2 years (Fig. 5A and B).

Discussion

The spine is not a common site for a GCTB, with a 2.5% incidence in the sacrum and 2.9% in the vertebrae above the sacrum. In the cervical spine, the incidence is extremely low and has been reported to be less than 1% in the literature. GCTB usually occurs after bone maturity, is slightly more prevalent in females, and is rare in the pediatric population.

The gross appearance of a GCTB is described as having a vascular, friable tissue with a surface that is reddish brown, yellowish brown, or yellowish gray. Based on histologic study, the red spots appear to be hemorrhagic, whereas the brown regions represent cellular stroma. Microscopically, there are moderately vascularized networks of round, oval, or spindle-shaped stromal cells and multinucleated giant cells that can be modified by hemorrhage and necrosis. Stromal cells, each containing a single large nucleus located in an indistinct cytoplasm, are loosely embedded in a sparse intercellular substance, and occasional mitosis may be observed. Multinucleated giant cells show pyknosis of their nuclei, and the number of nuclei in the giant cells varies greatly, with potentially as many as 50.

Fig. 4 (A) Histopathology showing the giant cell tumor. (B) High-power field of the histopathology showing the giant cell tumor.

Fig. 5 Follow-up computed tomography (CT) of craniocervical junction after 2 years shows no tumor recurrence. (A) CT scan sagittal cut; (B) CT scan axial cut at the C2 body level.
Differentiation between GCTBs and aneurysmal bone cysts (ABCs) may be challenging. A young age suggests ABCs, whereas an older age suggests GCTB.12 Lesions above the sacrum were once believed to be ABCs until proven otherwise.4 Histologically, both lesions contain giant cells, which make them difficult to differentiate. Cavernous vascular spaces are the hallmark of ABCs but are absent in GCTB.4,11 Microscopically, the giant cells in a GCTB lie among the stromal cells, which do not have intercellular spaces, whereas the giant cells in an ABC tend to be small with small nucleoli unlike the giant stromal cells in GCTB.4,11,12

It is not easy to differentiate GCTB radiologically, as it may be confused with ABC, brown tumor, osteoid osteoma, plasmacytoma, or even metastases, chordoma, and lymphoma of the spine.4,13

The radiographic features of a GCTB of the spine always start in the vertebral body with possible vertebral body collapse or extension into the nearby structures like intervertebral disk, adjacent vertebral body, spinal canal, or paraspinous soft tissues.14

The X-ray appearance of a GCTB in the spine, usually involving the body of the vertebra, is cystlike, with or without compression fractures of the involved vertebrae and with possible involvement of the pedicle or other portion of the arch of the vertebra and the body.4 CT is superior to conventional radiography and will outline the tumor extent, especially in the extraosseous portions, and reveal the bony integrity and stability.15,16

On MRI, GCTB may show heterogeneous or homogeneous signal intensity on T1-weighted images with possible areas of high-signal intensity caused by recent hemorrhage. The solid areas of the tumor have heterogeneous low to intermediate signal intensity on T2-weighted images. In more than 50% of cases, areas of low signal intensity may be exaggerated on T2-weighted, spin-echo images due to the presence of hemosiderin.11,13

ABCs may appear cystic, and trabeculae in the cyst may create a soap-bubble appearance in the lesion. The air-fluid levels may also account for the cystic component, and the lesions may be rim-enhanced with contrast on MRI.13,17

Because of its benign nature, the standard treatment is complete en bloc excision of the tumor, but the complex anatomy with proximity to vital neural and vascular structures in the cervical spine usually limits the en bloc removal of the lesion. A combined approach may be recommended to maximize the chance of tumor excision as well as the spine reconstruction and stabilization.18

In the presented case, the surgical team preferred to perform long occipitocervical fusion down to C5 to maximize the spinal fusion because the spine was posteriorly explored (cervical laminectomy) in the initial procedure that was done in the referring hospital. In addition, the pathology was uncertain (malignancy was still suspected), and there was no plan for another surgery at this stage. The anterior submandibular retropharyngeal approach for tumor resection and spinal fusion was planned after confirmation of the histopathology as GCTB. In retrospect, if the pathology was confirmed as a benign lesion, the occipitocervical arthrodesis in the pediatric age group should be limited to the minimum number of levels necessary to achieve satisfactory stabilization.

A surgical approach is always challenging with regard to the upper cervical spine. Moreover, decision making usually depends on the diagnosis, tumor type, extension of the lesion, presentation, and stability.3,7

A submandibular retropharyngeal approach provides excellent exposure to the anterior cervical spine from the craniovertebral junction up to C4 and facilitates reconstruction of the spine. In contrast, the transoral approach is considered only for biopsy and/or limited excisions of the lesion.7 The submandibular retropharyngeal approach has no connection with the oral cavity and allows placement of a graft to reconstruct or stabilize the spine with hardware at the same time, which is an advantage over the transoral approach. The disadvantages include the extensive dissection, prolonged exposure time, and a deep working field. In addition, a traction injury to the ninth and tenth cranial nerves and compression of the pharynx may cause transient deglutition impairment.7

Local recurrence is high when total resection is not achieved, especially when the tumor extends around the neural elements or adjacent vascular structures. The overall recurrence rate in the spine is between 25 and 45%.19 In the upper cervical spine, the recurrence rate is slightly higher because complete excision is not always feasible.

Radiation therapy as the primary treatment or as an adjuvant treatment has remained controversial. Some suggest complete excision must be attempted because of the benign nature of GCTB and because radiation therapy, which is considered for unresectable tumors and those with multi-level involvement, may increase the tendency for aggressive transformation of the residual tumor.8 However, some authors have had good results with primary radiation therapy followed by surgical excision,7 with no recurrence or malignant transformation at the latest follow-up.20

**Conclusions**

Giant cell tumor involving the second cervical vertebra in pediatric patient is uncommon. The lesion was surgically resected, and the spine was reconstructed using a combined posterior occipitocervical arthrodesis and anterior cervical retropharyngeal C2 corpectomy and fusion using fibular bone graft and fixation from C1 to C3, with no recurrence or eventful outcomes at the final follow-up 2 years after surgery.

The presented case was unique in terms of the tumor location, the patient’s age, and surgical management.

Disclosures
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