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Multilevel Giant Cell Tumor of the Cervical Spine: A Pediatric Case Report and Review of the Literature

Hammoud M*, Lakhdar F, Benzagmout M, Chakour K and Chaoui MF

Department of Neurosurgery, Hassan II University Hospital of Fez, University Sidi Mohammed Ben Abdellah, Fez, Morocco

***Corresponding author:**

Marouane Hammoud,
Department of Neurosurgery, Hassan II University Hospital of Fez, University Sidi Mohammed Ben Abdellah, Fez, Morocco,
E-mail: marouane.hammoud@gmail.com

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1. Abstract

Giant cell tumor of the bone is benign and expansile osteolytic lesions that occur infrequently in the spine above the sacrum. They are locally aggressive, with high rate of local recurrence.

We report a pediatric case of GCT in the cervical spine, revealed with cervical spinal cord compression,

We performed a complete excision associated with posterior cervical fusion. We decided not to administer radiotherapy to the patient after consulting with the radiation oncology department

At 1-year follow-up, the patient had shown no recurrence.

2. Introduction

GCT of bone are a rare locally aggressive lesions representing 4-10% of primary bone tumors [1], the spine location is even more rare and usually occurs in the sacrum [2].

The surgical management of these tumors, particularly when associated neurovascular compromise, is challenging in terms of achieving proper resection and spinal stabilization and ensuring no subsequent recurrence or failure of fixation.

Herein, we report a clinical case of a teenager with multilevel bone lesion of the cervical spine; total excision and posterior cervical fusion with C4-D1 laminar hooks fusion were performed.

Histological report confirmed the diagnosis of GCT, we decided not to complete by radiotherapy.

3. Case Report

A 14-year-old boy was admitted to our Neurosurgery department with 4 months history of cervical pain, 5 days before admission; the patient presented a rapid progressive tetraparesis, with increasing neck pain.

Physical examination found an overall limitation to his cervical mobility and hypoesthesia in right C6-C7 dermatomes

The muscle strength of the right upper and lower extremities was 2/5, and 4/5 in left upper and lower extremities, with a positive Babinski signs.

A spinal MRI shows the lesion displaying heterogeneous signal intensity with multiple cystic areas and fluid–fluid levels, involving the posterior elements, the right pedicles of C4 C5 C6. The lesion extends to the paravertebral soft tissue and spinal canal, causing spinal cord and nerve root compression. (Figure A, B).

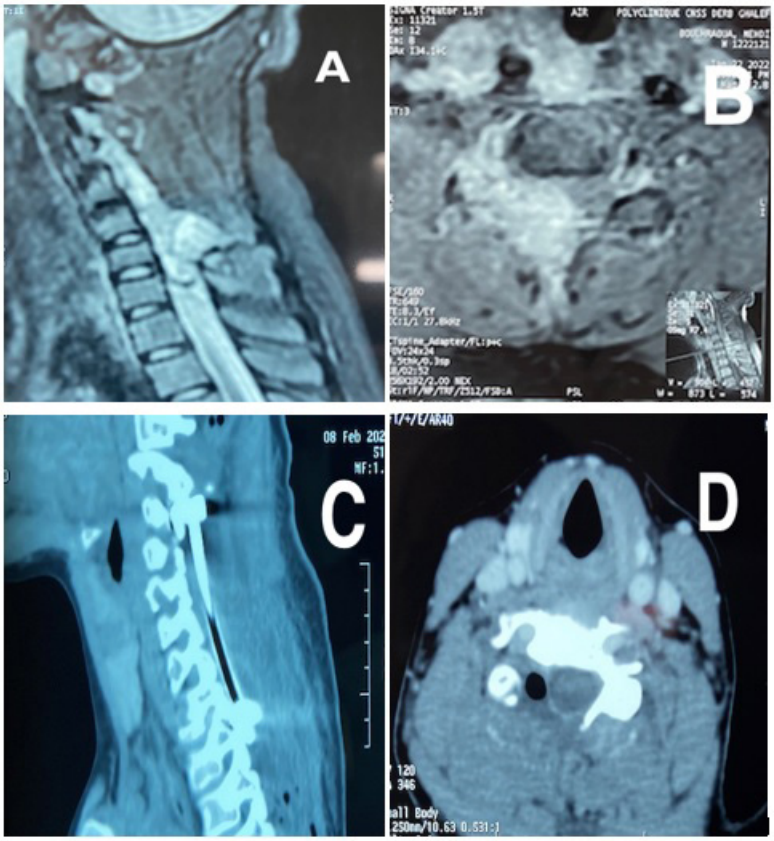
Because of the involvement of nerve roots, *en bloc* resection was not possible; we performed meticulous total excision; in a staged procedure followed by a posterior cervical fusion using C4-D1 laminar hooks to ensure spine stability (Figure C).

1 week after surgery, the patient was doing well, and has improved totally his neurological deficit.

Histological studies confirmed the diagnosis of GCT.

Radiotherapy was not given because of the proximity of the tumor to the spinal cord and the risk of radiation-induced myelopathy

After 1 year-follow-up, the cervical CT-scan control has shown no recurrence (Figure C, D).



Figures A: sagittal MRI of the cervical spine showing lesion extending from C4 –C7 along with mushrooming epidural tumor, causing severe cord compression.

B, axial MRI showing involvement of the posterior elements and nerve roots, with tumor extending into the transverse foramen.

C-D, Sagittal and axial CT Scan showing the final postoperative lateral view of the cervical spine after posterior approaches for excision of tumor and instrumental fusion.

4. Discussion

Giant-cell tumors of bone are rare, benign neoplasms involving the metaphysis of long bones. The incidence in the spinal cord varies between 1.4% and 9.4% and the sacrum is the most common spinal location [3]

Only 2 to 3% of GCT reported affect the spine above the sacrum [4]

The cervical location is even more exceptional; its incidence is less than 1%. [5,6].

There are only a few published reports focusing on GCT of the cervical in the literature. Nine case reports [7-15] and only one small series of 22 cases published by Junming et al [16].

GCT occurring in the cervical spine easily onsets between the second and fourth decades, and they are more common in female [16].

The most frequent symptoms of cervical spine GCT are pain in the neck and the shoulder and numbness in the extremities, GCT compressing the spinal cord can cause neurological deficit [8].

Histopathologically, giant cell tumor consists of abundant osteoclastic giant cells intermixed throughout the spindle cell stroma. Aneurysmal bone cyst-like areas are frequently detected in GCT, although solid areas in aneurismal bone cyst may be misdiagnosed as GCT. In Aneurysmal bone cyst, giant cells are smaller and giant cells are unevenly distributed [17].

On radiographs, spine GCT often shows an expansile osteolytic lesion. The lesion often produces collapse of the vertebral body, ranging from mild collapse to a complete vertebra plana [18,19].

On CT scans, the tumour has soft-tissue attenuation with no evidence of mineralized matrix. Areas of haemorrhage or necrosis may create a heterogeneous density with foci of low attenuation. The cortex at the lytic areas becomes thin or gets penetrated, or disappears with an associated soft-tissue mass [20].

From an anatomic point of view, in ¾ cases the tumor arises from the vertebral body and extends to the vertebral arch, The pedicle of the vertebral arch is always involved [19].

On MRI, the tumor has heterogeneous signal intensity on all sequences. It appears in hypo to iso signal on T2 weighted images. This feature is very helpful to distinguish GCT from other spinal neoplasms such as metastases, aneurysmal bone cyst, chordoma usually appears on hyper signal on T2 weighted images [21, 22].

GCT is usually well known to be locally aggressive tumor with benign character; it is associated with high rate of local recurrence.

The overall recurrence rate for spine tumors is 25 to 45% [18, 23].

Although, Some GCTs have a capacity for distant metastasis, Sanjay and Younge reported that metastasis of benign GCTs were slow-growing and had a good long-term prognosis [18].

Mondal et al. reported that factors regulating the local recurrence and metastatic potential of GCTs depend more on the tumors aggressiveness, than the histopathological appearance [24].

Thus, local control is a major prognostic factor for local recurrence [25].

The aim of the treatment is complete resection to prevent local recurrence while avoiding neurological structure damage and spinal instability [18, 26].

Various treatment options such as surgery, radiotherapy, embolization, cryotherapy, and chemical adjuvants are used for spinal GCTs [27].

Although, complete surgical removal is the best treatment, the rate of local recurrence is directly related to the quality of excision [28].

In the series of Campanacci et al. 27% which performed curettage inside the lesion had recurred versus none of the 58 cases that underwent radical excision relapsed [29].

In cervical spine location, because of the relatively complicated anatomic structure, radical excision is very difficult to realize in many cases, especially when the tumor was detected late and had extended to a wide range [28, 30].

In our case, based on preoperative imaging studies, the neurovascular and the peduncle involvement, we decided to perform an intralesional resection with cervical spine fusion.

Thus, we believe that aggressive surgical treatment for GCT is the best attitude because these tumors are locally invasive and have an unpredictable course. Although the treatment of recurrence, even if very aggressive, is always unsuccessful at long-term, Junming et al. suggested that the first time is the best opportunity to treat this disease [16].

Adjuvant Radiotherapy (RT) is an option to decrease postoperative recurrence in GCT, Especially, multilevel disease with residual tumor [25,31],

By now, debate continues on radiotherapy in GCT, the two principal's complications are the radiation myelopathy and the sarcoomatous transformation, therefore RT is reserved for recurrence cases [32].

In our case, after consulting with radiotherapy department, we decided not to irradiate the patient.

At 1-year follow-up, the patient has shown no recurrence.

5. Conclusion

Giant-cell tumors are rare, locally aggressive lesions that present a serious surgical challenge when they are located in the cervical spine. Although radical excision is the standard of treatment, however, it is not always possible to perform because of involvement of the anatomic structures such as vertebral arteries, nerve roots.

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