Bilateral C-1 lateral mass reconstruction following radical resection of a giant osteoblastoma of the atlas: case report

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Osteoblastoma is a rare, benign, osteoid-producing, and slow-growing primary bone tumor, typically arising in long bones or in the spine, with a slight male predominance. This report describes the surgical treatment of a giant C-1 (atlantal) osteoblastoma diagnosed in a young male patient with neurofibromatosis Type 1. The authors describe the clinical presentation, the surgical procedure for complete excision and stabilization, and results as of the 1-year follow-up. They detail a bilateral occipitoaxial spinal interarticular stabilization technique that they used after complete tumor excision. To the best of their knowledge, this is the first case of bilateral C-1 lateral mass reconstruction by this technique to be reported in the literature.

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KEY WORDS osteoblastoma; bilateral occipitoaxial spinal interarticular stabilization; C-1 lateral mass; neurofibromatosis Type 1; oncology

OSTEOLASTOMA is a rare, benign, osteoid-producing tumor, representing 1% of primary bone neoplasms.²⁹ It is more frequent in young males (mean age at presentation 20.4 years, range 6 months to 75 years; male/female ratio 2:1),¹ arising in the vertebral column in roughly 35% of cases.²⁵

Clinically, osteoblastoma is a slow-growing lesion whose nidus size of > 1.5 cm differentiates it from the smaller osteoid osteoma.²⁹ The most common clinical presentation is local pain.

For radiological diagnosis, CT scanning typically shows a bony lesion with cortical expansion and a radiolucent nidus. There can be surrounding bone rim sclerosis and multiple matrix calcifications, and more aggressive lesions can exhibit extensive bone destruction. MRI reveals soft-tissue extension, spinal cord or nerve root compression, and possible vascular compression due to tumor expansion.^{19,25}

Diagnosis is usually confirmed by biopsy, and the treatment of choice, for all tumor stages, is a wide en bloc resection when technically feasible as an inadequate tumor margin is the main factor that negatively affects the prog-

nosis.^{6,18} En bloc and marginal en bloc resection requires more extensive exposure to allow the surgeon access to the entire extent of the tumor.

In this report, we describe the treatment of an osteoblastoma diagnosed in a patient with known neurofibromatosis Type 1 (NF1). An association between osteoblastoma and NF1 has not been described and the relationship between the two is not certain in our case. We describe the surgical treatment of a giant osteoblastoma of C-1. Gross-total intralesional resection of the tumor was performed and followed by bilateral occipitoaxial spinal interarticular stabilization (bOASIS) for reconstruction. Both vertebral arteries were preserved. The OASIS technique following tumor resection has been previously described by our group for unilateral lesions.⁴

Case Report

History

A 12-year-old boy with known NF1 presented with a 6-month history of progressive neck pain. Initial investi-

ABBREVIATIONS bOASIS = bilateral occipitoaxial spinal interarticular stabilization; CVJ = craniovertebral junction; NF1 = neurofibromatosis Type 1; Oc = occiput; VA = vertebral artery.

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FIG. 1. A–C: Preoperative studies. Sagittal (A) and axial (C) CT scans showing destruction of the posterior arch and involvement of lateral masses of C-1. Coronal CT angiogram (B) showing displacement of the vertebral arteries. **D–F:** Sagittal T2-weighted (D) and T1-weighted (E) and axial T1-weighted Gd-enhanced (F) MR images showing a well-enhancing tumor with cord compression. Figure is available in color online only.

gation was done at an outside hospital with plain cervical radiographs, which were interpreted as normal. Cervical MRI, performed because of the patient's persistent neck pain, showed a mass centered on the posterior arch of C-1. The patient was then referred for surgical evaluation.

Examination

On physical examination the patient exhibited local tenderness in the C1–2 region at the nape of his neck and had limited neck movements, especially rotatory movements. General and neurological examinations showed normal findings.

Cervical CT scanning (Fig. 1A–C) revealed a giant ($4 \times 5.5 \times 2$ -cm) osteolytic tumor centered on the posterior arch of the atlas and extending into the lateral masses bilaterally. CT angiography showed that the tumor displaced the V₃ segment of the vertebral arteries (VAs) rostrally, although both remained patent. MRI confirmed the presence of a contrast-enhancing lesion; spinal cord compression was seen at the level of C-1 with a focal area of cord hyperintensity (Fig. 1E and F). The tumor extended into the posterior portion of the C-1 lateral masses bilaterally (Fig. 1C and F). Following multidisciplinary discussion, a mini-open biopsy was performed via a small midline posterior incision, and examination of the tissue confirmed the diagnosis of osteoblastoma. Because an en bloc resection was not considered to be technically feasible, a radical intralesional resection with reconstruction of the cranio-cervical junction was planned.

Operation

After induction of general anesthesia, the patient was

positioned prone on a Jackson table and his head was stabilized in a neutral position using a Mayfield clamp. Neurophysiological monitoring was used during positioning and throughout the surgery.

An extension of the previous midline posterior approach was performed to expose the suboccipital region down to C-2, including the posterior surface of the tumor. The mass was found to be firm and relatively avascular. Piecemeal intralesional radical excision was performed using the operating microscope. Both vertebral arteries (VAs) were identified during the resection and preserved, with a clear dissection plane between the tumor's capsule and the VA on either side. Portions of the tumor extending ventrally into the lateral masses of C-1 were resected completely using an ultrasonic bone scalpel, leaving only the anterior-most portion of the C-1 lateral masses (Fig. 2B). Following macroscopic gross-total resection of the tumor (Fig. 3), two appropriately sized titanium cages (Corridor, Globus Medical) were placed between the occipital condyles, whose convexities were drilled flat and cancellous bone exposed, and the superior facet surfaces of C-2 (Fig. 4). The cages were passed medial to the V₂ segment and inferior to the V₃ segment of the VA bilaterally such that no VA manipulation was necessary (see Fig. 4 for surgical details). Standard posterior instrumentation from the occiput (Oc) to the C-2 pedicles was then placed using image guidance (Fig. 4). A bone graft composed of autologous cancellous bone from the C-2 spinous process mixed with demineralized bone matrix putty (Grafton Putty, Osteotech, Inc.) was positioned inside and behind the cages bilaterally.

Somatosensory and motor evoked potentials remained normal throughout the procedure. Wound closure was performed in multiple layers in a standard fashion.

FIG. 2. Lateral view of the Oc–C1 joint, with the red line showing the “meridian” of the socket-ball joint. **A:** Normal Oc–C1 joint relation. **B:** Removal of the socket component of the joint. **C:** Downward and posterior slope of the condyle over the lateral mass. Copyright Iulia Peciu-Florianu. Published with permission. Figure is available in color online only.

Postoperative Course

The postoperative course was uneventful. The patient was mobilized the day after surgery without any cervical collar. The histopathological analysis confirmed the diagnosis of osteoblastoma. Postoperative radiography, CT, and MRI were performed during the patient's hospital stay and at the 6- and 12-month follow-up examinations (Figs. 3 and 5). These confirmed gross-total resection. At the 12-month follow-up, imaging demonstrated no tumor recurrence, and the patient remained pain free (0/10 on the visual analog scale) and neurologically intact.

Discussion

Osteoblastoma represents a rare benign osseous neoplasm, first described alongside its histological analog, the osteoid osteoma.^{15,20} Osteoblastoma predominantly affects young males in their 2nd or 3rd decade, being twice as

frequent in males as in females. Osteoblastoma affects the spine in 30%–40% of cases, predominantly at the cervical level,³³ arising in 85% of cases from the posterior elements, with a possible extension to the vertebral body.²⁵ It has also been described specifically at the atlantoaxial level in children, but it remains a rarity in the spectrum of primary bone tumors.²²

The association of osteoblastoma in a patient with known NF1 has not been described previously. To the best of our knowledge, this is the first described case of an osteoblastoma in a patient with NF1. This autosomal dominant neurocutaneous syndrome is associated with an increased incidence of a wide spectrum of benign and malignant tumors. There are reports of an association of osteosarcoma and NF1, due possibly to the loss of expression of neurofibromin, a product of the *NF1* gene with an important role in the activation of ras protein and in the proliferation and differentiation of cells.^{7,12} Although rare, in

FIG. 3. Postoperative 12-month follow-up MR images: sagittal T1-weighted (**A**) and T2-weighted (**B**) and axial T1-weighted Gd-enhanced (**C**) images showing complete resection and satisfactory decompression. Figure is available in color online only.

FIG. 4. Artist's view of the operative technique demonstrating initial tumor invasion (A), total excision with drilling of C-1 lateral masses (B), Oc–C2 cage positioning (C), and final result after positioning of the occipital plate and C-2 transpedicular screws (D). Copyright Iulia Peciu-Florianu. Published with permission. Figure is available in color online only.

non-NF1 patients, malignant transformation of osteoblastoma toward osteosarcoma has been described, especially after partial excision, and may be diagnosed years after the initial surgery.^{24,40} A subtype of osteoblastoma with aggressive features has been reported, with microscopic features that can blur the distinction with osteosarcoma.³⁵ The main criterion for distinguishing an aggressive osteoblastoma is the presence of epithelioid osteoblasts;⁵ it also has a propensity for local invasion and higher recurrence rates. These histological features were not found in our case.

From a surgical viewpoint, the main challenge for this case was adequate reconstruction following resection of the lateral masses of C-1. This was achieved with bOASIS reconstruction and occipitocervical fixation with subsequent bone fusion. We felt it necessary to reestablish the load-bearing capacity of the head with the use of cages following extensive bone resection beyond the “prime meridian” of the occipitoatlantal joints (see Fig. 2).

Functionally, the craniovertebral junction (CVJ) is designed for maximal capital mobility to allow panoramic visualization of our surrounding environment. The two contained motion segments have very different biomechanical properties and roles in head movement.³⁹ Stability is predicated primarily on intact ligaments, with osseous articulations designed for rotatory gliding motion between C-1 and C-2, and “hinge-like” motion between the Oc and C-1. The minimal bone constraints of these joints are subordinate to mobility requirements. Extensive literature exists regarding occipitoatlantal instability generated from condyle resection or tumor invasion.^{2,27,36} Evidence addressing instability generated by C-1 lateral mass tumor invasion or resection is lacking. The occipitoatlantal “ball and socket” joint allows flexion and ex-

FIG. 5. Postoperative studies obtained at 12 months. Sagittal (A), coronal (B), and axial (C) MR images showing the position of the intraarticular cages and transpedicular C-2 screws, with fusion visible at the site of the Oc–C2 cages (intraarticular graft and on-lay graft). Lateral (D) and anteroposterior (E) radiographs and 3D CT reconstruction (F) showing hardware and anatomy. Figure is available in color online only.

TABLE 1. Literature review of C-1 lateral mass reconstruction

Authors & Year	Approach			Cage	
	Dual Lat & Posterior	Posterior Only	VA Preserved	Unilat	Bilat
Wang et al., 2009		✓	✗	✓	
Chung et al., 2012	✓		✗	✓	
Winking, 2014	✓		✓	✓	
Bobinski et al., 2015		✓	✓	✓	
Present study		✓	✓		✓

tension, and the bony constraints resist translation in the sagittal plane. C-1 lateral mass resection beyond the meridian (Fig. 2A and B) removes the bony constraint of the “socket” and can result in the convex occipital condyle (ball) slipping posteriorly off the residual, down-sloping anterior portion socket of the C-1 lateral mass (Fig. 2C). Additionally, this bone resection likely renders the transverse ligament incompetent. The resultant loss of bone and ligamentous stabilizing constraints and loss of load-bearing capacity of the C-1 lateral mass requires reconstruction and poses very specific challenges for fixation and reconstruction techniques. The intact lateral masses of C-1 transmit the axial load from the occipital condyles through to the lateral masses of C-2.⁹ Moreover, ligamentous insertions on the lateral masses of C-1, primarily the transverse ligament, play a vital role in atlantoaxial stability and maintain correct articular surface alignment.^{11,26,34} Older techniques such as posterior wiring for CVJ instability effectively resist flexion and extension but poorly resist rotation and translation, which may lead to secondary failure in arthrodesis.³ Different posterior construct techniques have been described to counteract rotational and translation forces, such as transarticular screws.^{14,17} CVJ reconstruction after partial or complete excision of the C-1 lateral masses has been described only in case reports (Table 1).^{4,8,37,38} Small case series in which longer constructs were used in the subaxial cervical spine have also been published.^{9,13} What is unusual in our case is the extensive resection of both lateral masses of C-1, with loss of normal-load bearing capacity of the head. With regard to occipitocervical instability, reported case series and the biomechanics literature focus on the extent of occipital condyle resection,^{30,31,36} as well as on ligamentous integrity.^{10,23,32} Condyle-related instability is considered to be present with the development of an additional neurological deficit, a major deformity, and incapacitating mechanical pain after a condyle resection of greater than 75%.³⁰ Perez-Orribo et al. have shown that after an extensive resection of the condyle (i.e., > 75%) range of motion at Oc–C1 increases from $14.3^\circ \pm 2.7^\circ$ to $20.4^\circ \pm 5.2^\circ$ during flexion and from $6.7^\circ \pm 3.5^\circ$ to $10.8^\circ \pm 3.0^\circ$ during right axial rotation ($p < 0.01$), indicating the need for craniocervical fusion.²⁷ Complex reconstruction of the CVJ using expandable cages when a C-2 spondylectomy is performed has also been described, demonstrating the reestablishment of anterior weight-bearing capacity with cages between C-1 and C-2, in conjunction with poste-

rior constructs.¹⁶ Currently, biomechanical studies on reconstruction of C-1 lateral masses after destruction or resection are lacking. Nonetheless, in view of prior clinical studies describing reestablishment of load-bearing capacity of the head following C-2 spondylectomy,^{16,21,28} we thought it necessary in our case to reestablish head load-bearing capacity with addition of bOASIS cages in conjunction with a posterior construct, to enhance the likelihood of a durable arthrodesis.

Conclusions

In our case, we achieved a gross-total intralesional resection with a complete Oc–C2 fusion without any complications at the 1-year follow-up.

The described bOASIS technique is an option that can be considered when bilateral C-1 lateral masses have been resected and should be complemented by the placement of a posterior occipitocervical construct.

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Disclosures

Dr. Duff reports owning stock in KB Medical.

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Conception and design: Peciu-Florianu, Chittur Viswanathan, Barges-Coll, Castillo-Velázquez, Duff. Acquisition of data: Peciu-Florianu, Chittur Viswanathan, Zambelli, Duff. Analysis and interpretation of data: Peciu-Florianu, Chittur Viswanathan, Duff. Drafting the article: Peciu-Florianu, Chittur Viswanathan, Barges-Coll, Castillo-Velázquez, Duff. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Peciu-Florianu. Study supervision: Chittur Viswanathan, Barges-Coll, Duff.

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