Reconstruction of the C-1 lateral mass with a titanium expandable cage after resection of eosinophilic granuloma in an adult patient

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Spinal involvement occurs frequently in cases of eosinophilic granuloma (EG), but surgical treatment is limited primarily to those with spinal instability. Involvement of the cervical spine is rare, but primarily occurs in the vertebral bodies, and is normally amenable to anterior corpectomy and spinal reconstruction. The authors describe a 27-year-old man with pathologically proven EG who presented with complete destruction of the C-1 lateral mass requiring spinal stabilization. A titanium expandable cage was used to reconstruct the weight-bearing column from the occipital condyle to the superior articular surface of C-2 from a posterior approach, with preservation of the traversing vertebral artery. To the authors' knowledge, this is the first reported instance of reconstruction of the C-1 lateral mass using an expandable metal cage, which facilitated preservation of the vertebral artery.

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KEY WORDS reconstruction; expandable cage; eosinophilic granuloma; oncology

EOSINOPHILIC granuloma (EG) of the bone can present as unifocal or multifocal benign osseous destructive lesions. These lesions are in absence of cutaneous, pulmonary, hypophyseal, or other manifestations of extraskelatal involvement.¹² The incidence of EG within the adult population is difficult to ascertain due to the rarity and variability of disease presentation but has been estimated to be 1 case per 2–3 million people per year.¹⁶ In skeletal Langerhans cell histiocytosis (LCH), the most frequent site of involvement is the skull followed by the mandible, pelvis, and spine. Spinal LCH represents 6.5%–25% of all skeletal cases. The most commonly affected site is the thoracic vertebral body, whereas the least common location is the cervical spine.⁹

In this paper we present a case of EG involving the atlas in an adult who underwent resection of the lesion and craniocervical reconstruction of the lateral mass using an expandable titanium cage from a posterior approach, with preservation of the vertebral artery. To our knowledge, this is the first reported instance of using an expandable titanium cage to reconstruct the atlas.

Case Report

History and Examination

A 27-year-old man presented to the hospital after complaints of atraumatic sudden onset of severe upper cervical and occipital pain. His physical examination showed significant restriction of cervical range of motion in flexion, extension, rotation, and lateral bending, with pain at the initiation of movement. His neurological examination was normal, with normal motor and sensory evaluations.

Computed tomography of his cervical spine and skull base showed evidence of an expansile, lytic, intraosseous soft-tissue lesion with associated pathological fracture within the right lateral mass of the atlas, and rightward rotatory and anterior subluxation of the atlas relative to the axis. Lytic destruction was also noted within the right anterior occipital condyle (Fig. 1). Magnetic resonance imaging showed a 4.78 × 4.78 × 4.35-cm lesion that was T1 isointense and T2 mildly hyperintense. MultiHance contrast administration demonstrated heterogeneous enhancement localized to the periphery. Diagnostic cerebral

ABBREVIATIONS EG = eosinophilic granuloma; LCH = Langerhans cell histiocytosis.

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FIG. 1. Preoperative sagittal (left) and coronal (right) CT reconstructions showing destruction of the C-1 lateral mass as well as erosion into the occipital condyle (left).

angiography demonstrated co-dominant vertebral arteries with a 40% stenosis of the right vertebral artery at the level of the C-1 lateral mass and posterior lateral displacement consistent with tumor encasement, with mild to moderate tumor blush. Radiographic findings were suggestive of an aneurysmal bone cyst, giant cell tumor, lymphoma, or LHC. The metastatic workup results were benign, including contrasted CT scans of his chest, abdomen, and pelvis.

Several treatment options were considered, including percutaneous biopsy, open biopsy, and definitive resection and spinal reconstruction. Given his significant pain, as well as the degree of destruction of the atlas, an operative intervention was recommended to provide both pathological diagnosis and immediate craniocervical stability.

Operative Technique

Following fiberoptic intubation and induction of general anesthesia, the patient was placed into a Mayfield headholder and carefully placed prone. A midline incision was made from the inion to the level of the C-3 spinous process. The incision was carried down to the level of the dorsal fascia. The dorsal fascia was opened in the midline and the paraspinal muscles reflected from the skull base, the posterior arch of C-1, and the spinous process and lamina of C-2. We also exposed the spinous process and superior lamina of C-3. While taking care to preserve the C2–3 facet joint, we then carefully exposed the lateral mass of C-2. On direct visualization there did appear to be gross replacement of the right C-1 lateral mass with tumor.

Following exposure, stabilization of the occipitocervical spine was begun (VuePoint II OCT, NuVasive). An occipital keel plate was appropriately sized and modified so that it remained flush against the occiput. Occipital midline screws were placed with lengths predetermined based on measurements obtained from the preoperative CT scan. We then turned our attention to placement of the C-1 lateral mass screw on the left side only. In a subperiosteal fashion, the C-2 root was carefully dissected free from the caudal aspect of the C-1 lateral mass laterally. The lateral mass of C-1 was exposed in this fashion. Using the technique described by Goel et al.,⁸ a left C-1 32-mm lag screw was inserted. Next, we turned our attention to C-2 for insertion of bilateral C-2 pars screws. Using the technique described by Goel et al.,⁸ we inserted favored angled screws that were 3.5 mm in diameter and 22 mm in length.

A titanium hinged rod was cut to size and inserted with set screws on the left side, connecting the occipital plate, C-1 lateral mass screw, and C-2 pars screws. Satisfied with the interim stability, attention was then focused on resection of the mass. We began by skeletonizing the right vertebral artery along its entire length down the entrance into the superior aspect of the C-2 body. The vertebral artery was mobilized and protected via vessel loops (Fig. 2). With the C-1 lateral mass well separated from the vertebral artery, a midline cut was made using Kerrison rongeurs in the C-1 posterior arch. Using a combination of

FIG. 2. Operative illustration showing the position of the expandable cage between the occipital condyle and the C-2 superior articular surface, with the vertebral artery (red) reflected using a vessel loop. Copyright Neill M. Wright. Published with permission.

Kerrison rongeurs and a high-speed drill, the C-1 posterior arch was gently resected down into the lateral mass. At this point, the lateral mass appeared to be grossly replaced by tumor, which was purplish in color and soft in texture. The tumor was followed into the anterior aspect of the C-1 arch. This was confirmed under lateral fluoroscopic guidance with an instrument placed in the deepest point of our dissection. We could feel the thin remaining anterior cortex of the anterior arch itself. All visible tumor was resected. To facilitate further exposure, bipolar cautery and sternotomy scissors were used to ligate the C-2 nerve root at the dorsal root ganglion on the right side. This allowed a significantly larger corridor for visualization and resection of remaining tumor. Following resection, the superior articular facet of C-2 as well as the inferior aspect of the occipital condyle could be directly visualized.

As noted on the preoperative imaging, there was evidence of erosion into the anterior-inferior occipital condyle (Fig. 1). Using the high-speed drill, the occipital condyle was debrided down to healthy cortical bone. With the vertebral artery mobilized and protected, a titanium expandable cage (VBR, Ulrich Medical) was inserted between the inferior aspect of the condyle and the superior aspect of the superior facet of C-2. The cage was inserted under fluoroscopic guidance until appropriate purchase was achieved.

A titanium hinged rod was placed on the right side of the spine and secured with set screws. The occipital screws were tightened to the final torque. The C-1 screw on the left side was tightened to the final torque. We then compressed the C-1 screw against the C-2 screws to further seat the articular cage. The C-2 set screws were then tightened to the appropriate torque. At this point, the occipitocervical junction appeared quite stable (Fig. 3). The locking screw for the cage was then tightened to the final torque.

The wound was copiously irrigated with antibiotic-containing irrigation and hemostasis was obtained. The occiput, C-1 posterior arch, and C-2 spinous process and lamina on the left side were decorticated. A left-sided posterior arthrodesis was performed using tricalcium phosphate synthetic bone graft (Vitoss BA, Stryker) reconstituted with bone morphogenetic protein. A medium drain was placed and tunneled out through a separate stab incision. Five hundred milligrams of vancomycin powder was

inserted into the wound and subsequently closed in layers. The estimated intraoperative blood loss was 500 ml.

Postoperative Course

The postoperative course was uneventful. Following the procedure, the patient was monitored briefly in the postoperative anesthesia care unit and then transferred to the neurosurgical floor where he remained neurologically intact. His pain was managed via oral analgesia; he was sitting upright and ambulating out of bed on postoperative Day 2 with use of a cervical orthotic brace (Miami J). He was discharged home on postoperative Day 6 without complication. On postoperative Week 4 the patient received fractionated radiation with a total dose of 20 Gy delivered in 10 fractions of 2 Gy per fraction to the remainder of the disease at C-1 as well as the postoperative bed with margins. He tolerated the treatments well without complication. At his 3-month outpatient clinic evaluation he underwent both lateral and open-mouth radiographs (Fig. 3). At 1-year follow-up he underwent CT, which showed evidence of intact hardware with appropriate alignment and progression of bone fusion (Fig. 4). He also endorsed appreciable improvement in his neck pain at 1-year follow-up with an improvement of his visual analog scale score from 10 to 4.

Discussion

Eosinophilic granuloma is a benign osseous lesion that is a unifocal or multifocal form of LCH. It is a rare disease entity of unknown origin but primarily affects myeloid dendritic cells, lymphocytes, and macrophages mixed with eosinophils and neutrophils.⁴ EG of bone is a rare entity with an incidence of 1 per 200,000 per year among children and adolescents and 1 per 2–3 million among adults, with only 7.8% of these lesions localized to the spine.^{5,6,18} According to Wilner,²² the most commonly involved spinal segment is thoracic followed by lumbar and cervical at 54%, 35%, and 11%, respectively. In children, he demonstrated that 60% of their lesions involve C3–5, while in adults 54% demonstrated upper cervical involvement at C2–3. Of the 40 patients evaluated in this series, only 1 patient showed evidence of EG involving the C-1 vertebral body.

A common clinical feature of EG of the spine is pain, but children and adults can present differently. According to Bertram et al.,¹ 76% of children presented with pain complaints while 100% of adults endorsed pain. Restricted range of motion and/or torticollis was described in 76% of children and only 60% of adults. Among children and adults, 33% of patients with cervical lesions presented with neurological symptoms. According to Huang et al.,⁹ adult patients are more likely to present with neurological symptoms when compared with children (33% vs 67%, $p < 0.005$).

Current data suggest that children with immature bone and adults with mature bone may exhibit different disease courses that may affect treatment planning. Plasschaert et al.¹⁴ demonstrated no evidence of recurrence after 5 years in 17 children following biopsy and/or curettage and grafting. In contrast, 4 of 15 adult patients showed evidence of

FIG. 3. Postoperative lateral (left) and open-mouth (right) radiographs depicting the occipital-C1–2 construct with the expandable cage placed between the occipital condyle and the C-2 superior articular surface.

FIG. 4. Postoperative sagittal (left) and coronal (right) CT reconstructions at 1 year showing bone ingrowth through the cage.

recurrence at 2 years follow-up, requiring additional surgery. There are also data that support the notion that EG is a self-limiting process that shows evidence of spontaneous healing, and due to its propensity to avoid the vertebral body endplates and ossification centers, there is potential for reossification and reestablishment of some vertebral body height loss after a period of 6 months.^{3,17,19} Local therapy, including intralesional injection of corticosteroids, bracing, or casting, is recommended in clinical situations involving persistent pain. Operative intervention is reserved for patients with evidence of spinal instability or development of neurological deficits.¹⁶ Adjuvant therapies used to treat EG include chemotherapy, radiotherapy, or a combination of the two. Radiotherapy is recommended for patients with subtotal resections, or recurrent or disseminated forms of disease.¹¹ Radiotherapy is typically avoided in children due to the risk of radiation-induced side effects, including abnormal bone development due to growth plate injury. The LCH Histiocyte Society Evaluation and Treatment guidelines (2009) recommend systemic chemotherapy in multisystem disease.

With respect to our patient, a significant concern was the ability to reconstruct his anterior weight-bearing column while maintaining patency of his posterior circulation via his vertebral arteries. However, the craniocervical junction poses many unique challenges given its anatomy, close proximity to the brainstem and spinal cord, and allowance of limited view through narrow anteriorly projected corridors when approaching posteriorly. The 2 large lateral masses of the atlas transmit the majority of its load-bearing forces from the occipital condyles, superiorly, to the superior articular facet of C-2, inferiorly. Without an actual vertebral body the options for reconstruction are quite limited. There have been previously published accounts of using iliac crest and/or rib graft for lateral mass reconstruction in similar cases.^{2,7,13}

C-2 pars screws were chosen for 2 reasons. First, after resection of the C-1 lateral mass tumor, the superior, medial, and lateral borders of the pars interarticularis were directly visualized. Additionally, the C-1 lateral mass was destroyed by tumor, with likely subsequent increased rotational instability, and pars interarticularis screws have been shown to be more rigid in axial rotation in cases of occipitocervical instrumentation compared with C-2 laminar screws.²³

To our knowledge there has been only 1 prior report of using a titanium mesh cage for anterior column reconstruction following resection of an aneurysmal bone cyst at C-1.²⁰ Due to those authors' concerns for potential blood loss and risk of vertebral artery injury they opted to preoperatively sacrifice the vertebral artery endovascularly. They did note, however, that if they were unable to safely perform the vertebral artery sacrifice they would have attempted to skeletonize, mobilize, and transpose the vessel intraoperatively and attempt to preserve it.

Our decision to use a titanium expandable cage is multifaceted. The use of titanium expandable cages in surgical cases requiring reconstruction has already been extensively supported.^{10,15,21} The modular nature of the expandable cage allowed for optimal selection of its endplates for greater surface area and therefore bone contact, promoting fusion. The fully collapsed cage allowed for better visualization during insertion and minimized the risk of inadvertent injury to sensitive structures, such as the vertebral artery and brainstem. The unexpanded cage also allowed for direct visualization and confirmation of appropriate load bearing between the occipital condyle and lateral mass of C-2 during expansion and seating of the cage, allowing for a firm and secure fixation and reconstruction of the anterior column in conjunction with our posterior instrumentation. To further ensure stability, we compressed our posterior construct across the level of the expandable cage. To our knowledge, this is the first report of reconstruction of a lateral mass with use of an expandable titanium cage.

Our patient presented with intractable pain, as well as imaging that showed evidence of cranio-atlantoaxial instability necessitating resection of an EG of the lateral mass of C-1 and immediate reconstruction using a titanium expandable cage to restore stability to the craniocervical junction and provide symptom relief. This case represents a novel use of an expandable titanium cage to reconstruct the anterior column while preserving the vertebral artery.

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Disclosures

Dr. Wright has received royalties from NuVasive for the posterior cervical instrumentation system used in the patient but not for the cage that is the main point of the report.

Author Contributions

Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: Stephens. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors. Approved the final version of the manuscript on behalf of both authors: Wright. Statistical analysis: Stephens. Administrative/technical/material support: Wright. Study supervision: Wright.

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