En Bloc Resection of Cervical Sarcoma Involving C1: Report of Two Cases and Surgical Considerations

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Abstract

Study background: A two patient case series describing the surgical management of upper cervical sarcoma. Due to the density of critical neurovascular structures in the upper cervical spine, these rare sarcomas require primary surgical treatment that preempts local recurrence. Recurrence secondary to tumor spillage is problematic due to scar tissue formation and radiation effect creating surgically inaccessible tissue planes. En bloc resection of sarcomas during an index procedure provides the best chance at cure and prevention of local recurrence. Meticulous planning, familiarity with anatomy and surgical technique is critical for the success of these operations.

Methods: Two patients: a 30-year-old and 36-year-old female, were referred to our institution with malignant spine tumors involving C1. The first was found to have a left sided synovial sarcoma anterolateral to C1 and C2. The second presented with metastatic alveolar soft tissue sarcoma at C1. Both patients underwent multi-stage en bloc surgical removal of their tumors.

Results: Successful en bloc tumor excision and instrumented stabilization of the cervical spine without neurovascular complication was performed. Tumor margins were negative and x-rays demonstrated adequate spinal alignment. At six month followup, MRI evaluation demonstrated no local recurrence in either patient.

Conclusions: En bloc resection is a highly effective, but technically demanding method of treating upper cervical sarcomas. In conjunction with adjuvant radiotherapy, en bloc surgery has the lowest risk of local recurrence and highest quality of life outcomes. Due to the proximity of critical neurovascular structures in the upper cervical spine, meticulous planning, staging and technique is required. A multidisciplinary surgical team should be assembled that includes a head and neck, skull-base, neuro-endovascular and spine surgeon. With appropriate planning, understanding of anatomy and surgical technique, en bloc resections of upper cervical sarcomas can be successfully performed.

Keywords: Upper cervical spine; C1; Synovial sarcoma; Soft tissue sarcoma; en bloc resection; Vertebral artery ligation

Abbreviations: ICA: Internal Carotid Artery; MCA: Middle Cerebral Artery

Introduction

Sarcomas of the spine are uncommon; sarcomas of the upper cervical spine are exceedingly rare. Synovial sarcomas comprise 5-10% of all sarcomas and less than 1% of all malignancies. Few spinal synovial sarcomas have been reported [1-13]. Similarly, soft-tissue sarcomas localizing to the spine are also very rare [14-16].

In the absence of contraindications, en bloc surgical resection is the gold standard for treatment of spinal sarcomas [1]. A 2002 study on patients with primary spinal sarcomas by Talac et al demonstrated local recurrence rates of 11%, 33%, and 70% for patients undergoing en bloc resections, piecemeal resections, and all resections, respectively [17]. Despite diminished risk of local recurrence, en bloc resection of upper cervical lesions is complicated by the density and proximity of important anatomical structures such as the vertebral arteries, carotid arteries, jugular veins, esophagus, trachea and lower cranial nerves. Extensive neck dissection, arterial ligation and nerve root sacrifice are often required in order to mobilize tumors away from vital structures [18,19]. Consequently, sarcoma surgery in the atlantoaxial region requires a multidisciplinary, staged approach with extensive use of operating room technology. Navigation-guided biopsy, spinal and neuroangiography, intraoperative cranial nerve EMG monitoring, high resolution imaging and experienced ICU care are required for planning, execution and recovery. The coordinated cooperation of specialists in head and neck, skull-base, neuroendovascular and spine surgery is essential. Here we describe two successful en bloc resections of atlantoaxial sarcomas and the knowledge acquired in planning, staging and surgical anatomy.

Cases

Case 1

A 30-year-old female presented to our clinic with a left-sided neck mass and increasing pain of 2 months duration. Past medical history was significant only for migraine headaches. Neurologic assessment was without abnormal findings. Imaging studies, including a CT scan and an MRI of the neck, revealed a 2 x 4 x 4 cm mass encasing the left vertebral artery extending from the lateral aspect of the C1 lateral mass distally to the C3-C4 facet (Figure 1A-B). PET scan, which was done to rule out metastatic disease, found clinically significant uptake only in the cervical lesion (Figure 2). Needle biopsy was performed and the results were most consistent with Ewing's sarcoma.

The patient was initially treated with neoadjuvant chemotherapy. Post-chemotherapy MRI revealed no evidence of tumor regression (Figure 3A-B). The patient was offered further options for local control including surgery and radiation therapy. She decided to proceed with en bloc surgery in order to minimize local recurrence and decrease the likelihood of post-radiation tumorigenesis. Due to the encasement of

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Figure 1: a) Axial CT scan slices demonstrating bony erosion on the left lateral aspects of C2 and C3 with a visible soft tissue mass. b) Axial T2 MRI slices at the C2 and C3 levels demonstrate tumor encasement around the left vertebral artery and internal carotid artery.

Figure 2: PET revealed no evidence of metastatic disease. The left cervical region demonstrated increased uptake of 18 F-FDG.

Figure 3: a) Pre-neoadjuvant chemotherapy sagittal T2 MRI sequence image showing tumor volume. b) Post chemotherapy treatment sagittal T2 MRI sequence image showing no apparent evidence of tumor regression.
the left vertebral artery and the need to ligate and sacrifice it, the patient was first evaluated by a neuro-endovascular surgeon. She underwent a balloon test occlusion of the left vertebral artery (which was tolerated) followed by coil-assisted embolization (Figure 4). The patient was scheduled to undergo an initial posterior approach with left vertebral artery sacrifice. This was to be followed by a test occlusion of the left ICA at a later date in anticipation of a final anterolateral approach with ICA sacrifice and en bloc removal of the tumor mass.

**Operative procedure: Stage 1:** The patient was positioned prone in a Mayfield head-clamp. Posterior midline incision was made and subperiosteal dissection of the paraspinal muscles was performed. Synthes posterior cervical instrumentation was placed on the right: a C1 lateral mass screw, a C2 pedicle screw and a C3 lateral mass screw. Decompressive left-sided hemilaminectomies at C1, C2 and C3 were performed and the C2 and C3 nerve roots were transected. The left vertebral artery was then ligated. A partial vertebrectomy was performed with sagittal cuts through the anterior columns of C1 and C2 on the left (Figure 5). The patient tolerated the surgery without complication and was admitted to the ICU.

**Stage 2:** In anticipation of the next stage of surgery requiring ICA sacrifice, a balloon test occlusion of the left ICA was performed. While some cross-over flow from the anterior communicating artery into the left middle cerebral artery distribution was noted, it was insufficient to prevent onset of contralateral motor deficits (Figure 6). Consequently, in order to bolster blood flow into the left anterior circulation, a low-flow superficial temporal artery to MCA bypass was performed. Post-bypass catheter angiogram and CT-angiogram demonstrated incomplete filling of the left MCA from the donor vessel (Figure 7). The patient was closely monitored in the ICU for signs of neurological deficit. It was determined that, despite the risk of leaving tumor behind, the left ICA had to be spared and the decision was made to proceed with the final stage of surgery.

**Stage 3:** With the assistance of head and neck surgery, the patient was positioned in right lateral decubitus position for a left-sided far lateral approach to the tumor. Transdermal electrodes for continuous EMG monitoring of the facial, glossopharyngeal, vagus, hypoglossal...
and spinal accessory nerves were placed and EMG responses verified. A horizontal incision made laterally at the level of the antitragus running across the occiput and temporal bone was joined to the previous midline incision. An occipito-cervical musculocutaneous flap was raised in a subperiosteal fashion. The previous cuts through C1 and C2 were identified and further dissection was performed to mobilize the tumor in the anteromedial, caudal, lateral and cephalad directions. The jugular vein was identified and it, along with the ICA, were mobilized laterally as the dissection proceeded along the anterior and cranial aspect of the tumor. The soft tissues around the tumor were divided and the tumor was excised en bloc. Frozen section analysis of the tumor suggested synovial sarcoma rather than Ewing’s sarcoma. Frozen section analysis of the margins was negative with some suspicion of tumor cells along the posterior margin. The wound was closed in layers and a hemovac drain was placed.

**Postoperative course:** The patient was taken to the ICU where she was monitored. Mild right-sided sensory and motor deficits were noted that resolved by the time of discharge. She was discharged home on postoperative day five in a Miami-J collar with instructions to follow-up in clinic.

**Case 2**

A 36 year old female with a history of metastatic alveolar soft tissue sarcoma presented to clinic with increasing neck pain secondary to a known C1 mass that had been previously treated with external beam radiation. She had also undergone surgical treatment for tumor deposits in her pelvis, humerus and lungs. Her neurologic exam upon presentation revealed no findings. MRI with and without contrast of the cervical spine was performed demonstrating interval growth of the C1 mass with encasement of the left V3 segment of the vertebral artery (Figure 8A-B). The patient no longer desired non-operative treatment and opted for surgical management. As a prologue to ligation, she underwent balloon test occlusion of her left vertebral artery followed by endovascular occlusion, which was well tolerated (Figure 9A-B).

**Operative procedure: Stage 1:** As in case one, the patient was positioned prone and underwent subperiosteal exposure. Synthes posterior spinal instrumentation was placed with an occipital plate, right-sided C1 lateral mass screw and C2 pedicle screw (Figure 10). Left hemilaminectomies were performed from C1 to C3. The C1 and C2 nerve roots were transected and the left vertebral artery was ligated. A partial vertebrectomy of C2 was performed and a silastic sheet was placed lateral to the spinal cord and medial to the C1 lateral mass to prevent tumor spillage. Due to erosion of the C1 lateral mass, the vertebrectomy was planned for the next stage via an anterolateral approach. The patient tolerated surgery well and was admitted to the ICU for observation.

**Stage 2:** On postoperative day eight, the patient underwent the second stage of surgery. In conjunction with a skull-base and head and neck surgeon, a far lateral approach to the cervical mass was performed. The lower cranial nerves were again monitored with EMG. The patient was positioned right lateral decubitus and a musculocutaneous flap was raised (Figure 11A-C). The internal carotid artery, digastic and sternocleidomastoid muscles, facial, vagus, spinal accessory and hypoglossal nerves were mobilized and the skull base dissected free of

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**Figure 8:** a) Axial T1 post-contrast images at the C1 and C2 reveal an avidly contrasting mass infiltrating into the left occipital condyle and C1 lateral mass. b) Coronal T2-weighted slices demonstrate encasement of the V3 segment of the left vertebral artery.

**Figure 9:** a) Pre-embolization left vertebral artery selective catheterization reveals V3 segment tortuosity due to compression from the surrounding tumor. b) Post-embolization occlusion of the vertebral artery.
muscle and tissue attachment. The tip of the mastoid and supracondylar bone was drilled away to improve visualization. At this point, our spinal surgery team entered into the operation. The C2 vertebrectomy was completed with an osteotome directed posterior to anterior. The C1 vertebrectomy was performed in a similar fashion across the anterior ring. The occipital condyle was finally removed with a burr and osteotome thus mobilizing the tumor medially, anteriorly, superiorly, laterally, and inferiorly. The tumor was removed entirely in an en bloc fashion (Figure 12 and 13). The wound was closed in layers and a deep drain was placed.

Postoperative course: The patient was monitored in the ICU. No postoperative complications were noted. On postoperative day 4, a halo was placed for stabilization due to removal of her condyle in the absence of bilateral fixation. She was discharge home on postoperative day six with clinic follow up.

Discussion

Histology, anatomy, location, systemic considerations and disease burden are all important considerations in the management of primary spinal tumors. Management can include radiation therapy, chemotherapy and surgery. Most spinal tumors, however, are treated through combinations of these modalities rather than the exclusive use of any one method [18-20]. Treatment algorithms are lacking for most spinal tumor histologies due to their rarity, a dearth of well-controlled studies and reliance on traditional treatment methods.

Radiation therapy is often used to treat primary spinal tumors in lieu of surgery, however, its administration is fraught with the risk of radiation-induced myelitis, secondary cancers, tissue fibrosis, hypothyroidism, infertility and a host of other complications – both acute and chronic [17,18,21-23]. The relative radio-resistance of many spinal tumors, especially sarcomas, requires high dose radiation protocols often approaching 50-60 grey fractions – the threshold at which radiation myelitis develops [21,22].

Neoadjuvant chemotherapy is often used in soft tissue sarcomas located near neurovascular structures to decrease the size of the tumor [24]. However, surgery remains the definitive final treatment pathway in patients who receive chemotherapy.

While surgery is the mainstay for treatment of primary malignant spinal tumors, not all surgical methods are equal in benefit or efficacy. The existing literature has documented en bloc surgical resection as providing the best chance for cure and prevention of local recurrence of spinal tumors [25-32]. The prevention of local recurrence is a critical consideration in the treatment of upper cervical sarcomas due to the density of critical anatomy in this region as well as the variable response of spinal sarcomas to chemotherapy and radiation. Boriani et al, in a study of 22 patients with spinal chondrosarcoma, demonstrated a local...
En bloc resection of sarcomas or spinal tumors involving the C1 and C2 level are challenging surgeries that push the envelope of surgical planning, management and execution. With the requisite armamentarium in the form of hospital resources, technology, coordination, knowledge and skilled specialists, these surgeries can be performed with substantial benefit to patients.

References


