

Clinical Image

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Adolescent presenting with bilateral neck and shoulder pain

Clinical Presentation

An 18-year-old girl presented with a 6-month history of slight bilateral neck and shoulder pain. The pain was constant and exacerbated when abducting her upper extremity. The pain was described as better at night. Medications, including aspirin, offered little relief. A hard unmoveable mass could be felt in the cervical back, approximately 10x8 cm in size. The neurological examination was normal with the exception of dysesthesia in the C5 dermatome. Routine serologic tests were all normal. Her family and medical history were unremarkable. Preoperative plain radiographs showed the osteolytic lesion located in the vertebral body and posterior elements of C5 with pathological anterolisthesis at the C4 level. The computed tomography (CT) scan of the cervical spine demonstrated a large expansile multiseptated mass, with a size of approximately 7.0x5.5x3.0 cm, containing fluid-fluid levels, involving the posterior elements, the pedicle, and the vertebral body of C5, and causing effacement of the C4/5 neural foramen. There was compression of the spinal cord mainly at the level of C5. Fortunately, the vertebral artery is not encased by the disease process. Magnetic resonance imaging (MRI) of the spine demonstrated a local destructive tumor lesion of the fifth cervical vertebrae with compression of the spinal cord and paravertebral involvement (Figure 1).

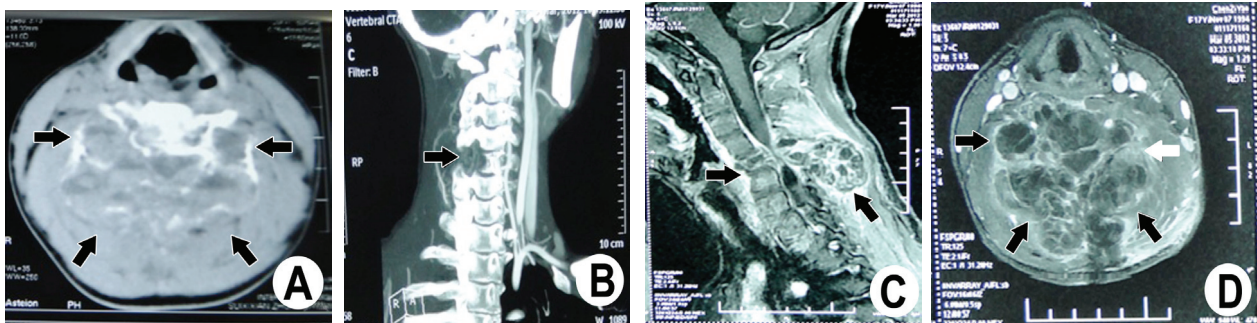


Figure 1 - Photograph showing A & B) Computerized tomography scan of the cervical spine showing a large expansile multiseptated mass, containing fluid-fluid levels, involving the posterior elements, the pedicle, and the vertebral body of C5. C & D) Magnetic resonance at the level of C5, demonstrating a cystic lesion with fluid-on-fluid levels with extension into the spinal canal.

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Questions

1. What is the diagnosis?
2. What is the etiology of this condition?
3. What is the management?

Answers

1. Due to the typical image performance, a diagnosis of an aneurysms bone cyst (ABC) variant with an unknown secondary process was considered. According to the Weinstein-Boriani-Biagini (WBB) staging system, the extension of the lesion occupied all the sectors except the eighth, and the layers A to D. The CT-guided needle biopsy suggested it was a secondary ABC on the basis of an osteoblastoma, which was confirmed by surgical pathology (Figures 2A-2C).
2. ABC associated with another tumor is referred to as a secondary ABC. The histogenesis of ABC remains unclear. Current theory holds that ABC is a nonneoplastic reactive condition, which is aggressive in its ability to destroy and expand bone. The osteoblastoma initiates an osseous arteriovenous malformation. With the change in hemodynamics, the vein pressure is elevated, the vascular bed expands, and the bones are destructed, which leads to the ABC.³
3. Surgery was performed after the selective arterial embolization. We resected the tumor through an anterior-posterior approach and then reconstructed the spine by anterior-posterior spinal instrumentation. Anterior surgery was performed by a standard left side approach to expose the C4-C6 levels. She was positioned supine on a radiolucent table with the head placed in a Mayfield pin headrest. A Caspar-Distractor was used to distract the C4/5 and C5/6 disk spaces. Discectomy of C4/C5 and C5/6 disk were carried out in sequence under microscopic visualization. Next, cleaning of the endplates of C4 and C6 with curette was performed. Then, the vertebral body of C5 was removed with the posterior longitudinal ligament, leaving only the most anterior lateral bony coverage of the vertebral artery on both sides. Finally, the posterior longitudinal ligament was removed, and the spinal canal was decompressed. A titanium mesh of suitable size was implanted. A cervical plate of suitable length was fixated with anglestable screws. After the anterior operation, she was turned over and underwent the C6 laminectomy, C5/6, and a C6/T7 facetectomy were also removed. Tumor resection was followed by a C3-C7 posterolateral instrumented fusion. Total blood loss was 2600 mL. She was completely disease free after surgery, no recurrence was observed. Plain film showed implants were well placed after 2 years follow up (Figure 2D).

Discussion

Osteoblastoma is a rare bone tumor with a reported incidence between 10-25% of primary osseous spine tumors. Ten percent of all osteoblastomas exhibit secondary changes consistent with ABC. However, this combination in the cervical spine is rare.^{1,2} Of the radiology techniques, CT is the preferred imaging modality

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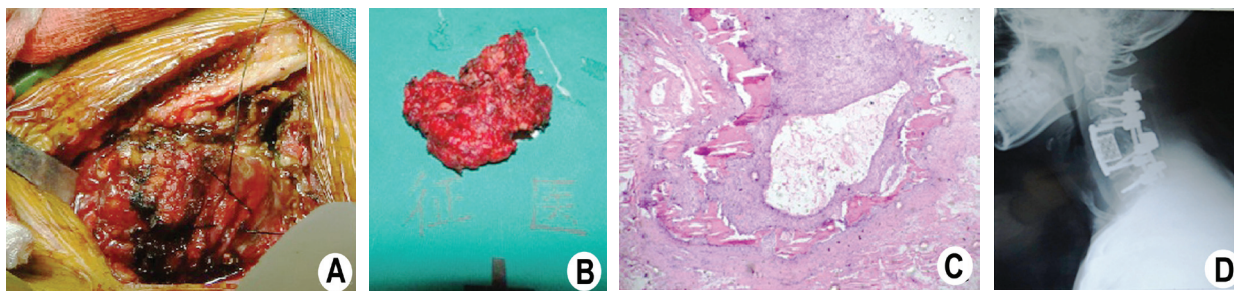


Figure 2 - Photograph showing A-C) In gross, the excised lesion was 7.0x5.5x3.0 cm in size and showed evidence of cystic blood-filled spaces. On histological examination, the lesion was well circumscribed with a sclerotic bony rim and contained haphazardly arranged bony trabeculae surrounded by small spindle-shaped cells and few multinucleated osteoclasts, thin-walled blood vessels fill the intervening stroma. D) Postoperative x-ray of the cervical spine showing the proper alignment of the spine.

in indentifying osteoblastoma, since it can identify the lesion, degree of sclerosis, and extent of bony involvement. While MRI can clearly revealed ABC with a fluid-fluid interface with hypointense signals on T1-weighted imaging and hyperintense signals on T2-weighted imaging with contrast enhancement of the septa.^{3,4} Osteoblastoma and ABC of the spine, while considered a benign tumor, is locally aggressive. Neither radiotherapy nor chemotherapy has a definite therapeutic effect on this lesion. Postirradiation sarcoma has been reported in the management of osteoblastoma, which makes this method of treatment inappropriate. Once a diagnosis is established they should be treated with gross total resection to minimize the risk of recurrence and potential malignant degeneration. The appropriate surgical treatment goal for osteoblastoma is en bloc excision of the lesion. “En bloc” indicates an attempt to remove the whole tumor in one piece, together with a layer of healthy tissue. Stener⁵ provide a detailed report firstly, and Tomita et al⁵ systemized the procedure. Studies shows recurrence rates of approximately 50% on the subtotal resection and 10% local recurrence to those patients with complete excision. However, this was not possible to perform on our patient because of her WBB stage. Both osteoblastoma and ABC are very vascular tumors, in order to permit a more aggressive surgical approach, the use of selective arterial embolization is recommended. As it facilitates resection by reducing intraoperative blood loss. Embolization also has the potential to improve outcome in patients with spinal tumors.⁴ We presented the largest complete excised case to date with microsurgical instrument. The satisfactory follow up prompted us to conclude that the appropriate treatment is a complete excision of the lesion if anatomically possible. However, considering the rich vascular supply of ABC and the complicated anatomic relationship of cervical spine, it might not always be possible to remove the entire lesion.

In conclusion, the technique of cervical total spondylectomy carries greater risks and requires more attention to the operation indication. We recommend the use of selective arterial embolization as it facilitates resection by reducing intraoperative blood loss, then a complete excision of the osteoblastoma with secondary ABC should be followed up.

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