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Case Report

Aggressive osteoblastoma involving the craniovertebral junction: A case report and review of literature

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Abstract

Osteoblastoma (OB) is a rare bony neoplasm constituting around 1% of all primary bone tumors. Although the vertebrae and long bones are the most common sites affected by OB, skull remains a relatively uncommon site of occurrence. Aggressive variant of OB is histologically intermediate between an indolent conventional OB and a malignant osteosarcoma. To the best of our knowledge, aggressive osteoblastoma (AO) affecting the craniovertebral junction has not been previously described in the literature. In this report, we present a 34-year-old gentleman who presented with a mass involving the left side of the neck and oral cavity along with ipsilateral lower cranial nerve paresis. Computed tomography and magnetic resonance imaging scans of the craniovertebral junction revealed a heterogeneously enhancing expansile lesion with areas of destruction involving the clivus, left sided jugular foramen and left side of first two cervical vertebras. Angiography showed distortion of the V3 segment of the left vertebral artery and shift of the ipsilateral internal carotid artery. The tumor was maximally excised through far lateral approach. Histopathologic examination revealed a diagnosis of AO. The patient was referred for radiotherapy for the residual tumor and was doing well at 5 months follow-up.

Key words: Aggressive, epithelioid, osteoblastoma, pathology, skull base, surgery

INTRODUCTION

Osteoblastoma (OB) was first described in the English literature by Jaffe and Mayer in 1932.[1] In the year 1956, Jaffe and Lichtenstein independently proposed the term "benign OB" to identify an osteoblastic osteoid-forming lesion similar to osteoid osteoma (OO), but having a greater growth potential.[2,3] Some of these seemingly innocuous tumors do, however, tend to have local destructive growth pattern and recurrence. Dorfman labeled these variants as aggressive osteoblastoma (AO)[4] and suggested that the presence of epithelioid osteoblasts was a distinctive histologic feature in these variants.[5]

OBs are very rare and constitute about 1% of all primary bony tumors. These tumors tend to involve long bones and vertebral column.[6] OB is rarely known to involve the skull and when they do, mandible followed by the fronto-temporal calvarial regions tend to be the most frequently reported sites.[7-10] Although, there are reports of involvement of skull base like the sphenoid sinus,[11] we are not aware of any reports of involvement of the craniovertebral junction by the aggressive variant of this tumor.

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Herein, we report the clinico-radiological and pathological findings of AO affecting the craniovertebral junction in a 34-year-old man and present a review of the literature.

CASE REPORT

The present case report is about a 34-year-old gentleman who presented with a painful swelling behind the angle of left mandible of 5 months duration. He also complained of change in character of voice, dysphagia and weakness of left upper limb for 3 months prior to admission.

On neurologic examination, he had IX, X and XI cranial nerve palsy on the left side. There was a 3.5 cm × 3.0 cm bony hard swelling underneath the angle of left mandible leading to obliteration of post auricular groove. It was extremely tender. On examination of the oral cavity, the swelling was bulging into the tonsillar fossa and compressing the palatal arches.

Since, the mass was palpable externally, fine-needle aspiration was attempted pre-operatively (twice) and on both occasions, but it was found to be inconclusive.

The axial and reformatted sagittal computed tomography showed a large expansile and destructive bony lesion involving the foramen magnum on the left side from lower clivus up to the axis. There were calcifications seen within the tumor. Thin sclerotic rim was visible around most of the periphery of the lesion [Figure 1a-d]. The lesion had involved the lateral masses and part of anterior and posterior elements of upper 2 cervical vertebrae.

Magnetic resonance imaging showed a heterogenous mass (size approximately 5.3 cm × 6.2 cm × 5.5 cm) with strong post contrast enhancement. The lesion was displacing left internal carotid artery peripherally causing mild compression and had involved the left vertebral artery, the oblique part of V3 segment [Figure 2a-d].

With pre-operative impression of a high grade bony lesion, the patient was planned for surgical decompression of the mass through left sided far lateral approach. At surgery, near total excision of the tumor was achieved, deliberately leaving a thin rim of tumor tissue attached to posterior pharyngeal wall to prevent opening the oral cavity [Figure 2e and f]. The tumor was very vascular and bled profusely. Occipito-cervical fusion was also done. The patient recovered uneventfully after surgery. Histopathological evaluation of the resected tissue showed epithelioid osteoblasts lining the bony trabeculae separated from the thin walled vessels by the osteoclastic giant cells. Mitotic figures were also seen. Hence, a pathological diagnosis

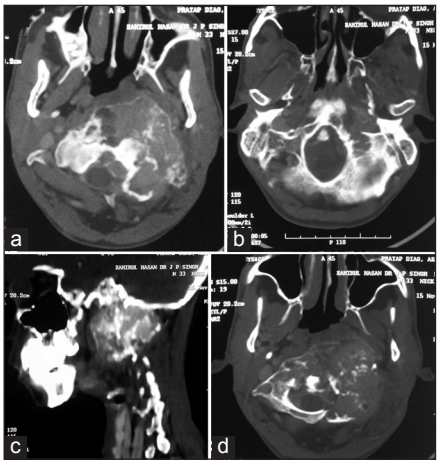

Figure 1: (a-d) Computed tomography scan of the craniovertebral junction shows expansile mass involving the clivus, occipital squama including the jugular foramen on the left side. There is involvement of the left sided elements of C1 and C2 vertebrae. Vertebral foramen of C1 is obliterated on the left side and anterior arch is involved more extensively than the posterior one. Internal calcifications can be seen and a thin peripheral bony rim can be seen around the tumor, which is absent at places

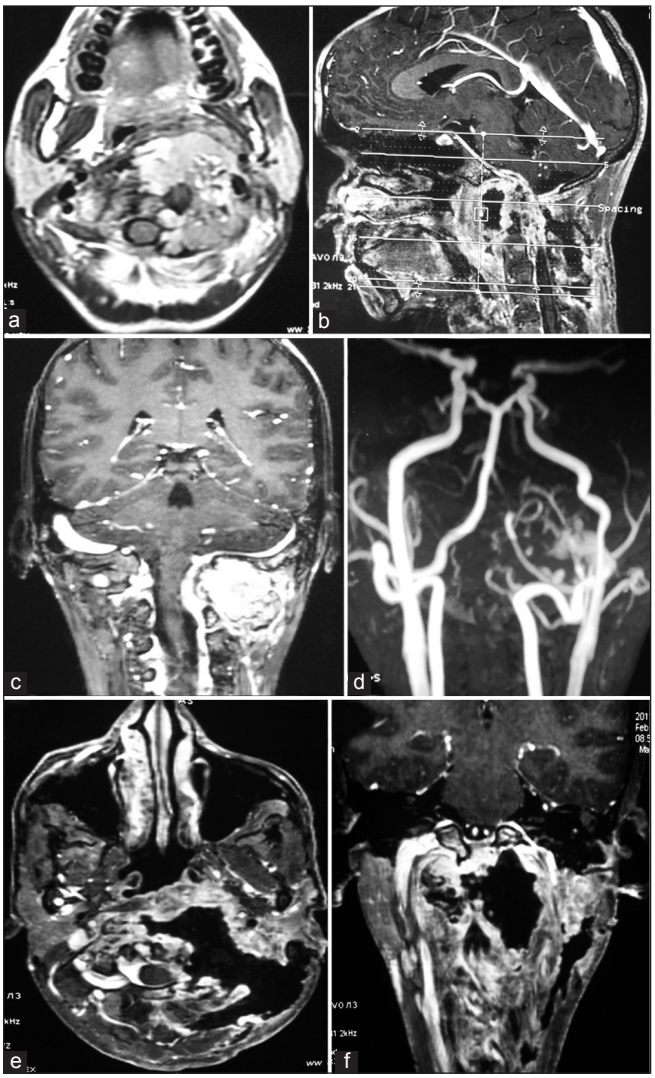

Figure 2: (a-c) The tumor is heterogeneously but avidly enhancing and extending inside the spinal canal but no significant neuraxial compression is seen. Mass is anteriorly pushing the oral cavity and nearly obliterating it on the left side. (d) Compression and thinning of ipsilateral vertebral artery (the oblique segment of V3) and displacement of internal carotid artery. (e and f) Post-operative images show surgical cavity with residual enhancement anterolaterally

of AO was made [Figure 3a and b]. The patient was referred to radiotherapy for residual lesion. At 5 months follow-up, the patient was doing well.

DISCUSSION

OBs are rare primary bony tumors constituting only 1% of all cases.[6] These were first described in the English literature by Jaffe and Mayer in 1932.[1] In the year 1956, Jaffe and Lichtenstein independently proposed the term "benign OB" to identify an osteoblastic osteoid-forming lesion similar to OO, but having a greater growth potential.[2,3] It most commonly affects males during the second decade and often arises within the vertebral column and long bones.[6] Approximately 10-12% of OB occur in the maxillofacial skeleton, especially the mandible.[6,7] Skull base is an extremely uncommon site of OBs.[11]

The pathological spectrum of osteoid forming primary bone tumors includes OO and OB at one end and osteosarcoma (OS) on the other. OO and OB differ only in size, potential for progression and propensity to produce extreme pain. Nevertheless both are benign and respond well to surgery. On the other side, OS is known to be malignant with frequent recurrences and metastasis in spite of seemingly adequate surgery. There are, however, some tumors with features nearly similar to the conventional OBs which tend to display aggressive behavior with recurrences and a locally destructive growth pattern. Dorfman examined one such case of his and 23 others which were reported until then and along with Weiss, named them AO.[4] They suggested that the presence of epithelioid osteoblasts was a distinctive histologic feature.[5]

AO have some subtle histopathological markers that need to be specifically looked for. These include presence of large epithelioid osteoblasts lining the bony trabeculae, presence of a moderate number of mitotic figures and invasion into the surrounding bone and soft-tissue.[5,12-14] Conventional OBs are lined by osteoblasts and lack any mitotic figures and invasion into surrounding structures. In contrast, OSs have abundant osteoid, have abundant mitotic figures and have areas of necrosis.

Due to the relative rarity, the incidence and distribution of AO are currently unknown.[15] Reports of skull base AO are exceptionally rare.[11] Both OB and AO often present with local pain and swelling; however, AO tends to affect an older age group than OB, usually arising in the third or fourth decade.[6] The radiographic appearance of AO is similar to OB, consisting of a circumscribed lytic defect sometimes surrounded by a sclerotic rim, although a more aggressive appearance including significant cortical expansion and destruction can be seen like in our patient. Intralesional radiopacities of varying amount and density have been described.[6,16] OBs at certain locations like spinal canal and skull base, irrespective of whether they contain epithelioid cells or not, would have aggressive clinical behavior. In these cases, the clinical aggressiveness appears more dependent on the precise location and size of the tumor

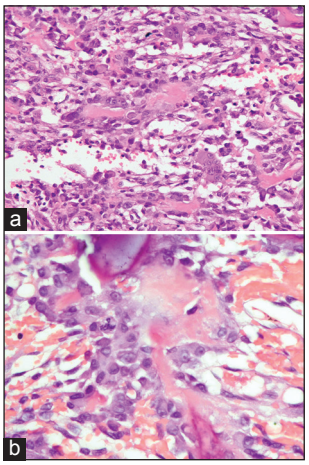

Figure 3: (a) Irregularly laid down osteoid rimmed by epithelioid osteoblasts (single arrow) separated by thin walled vascular channels and multinucleate ostoclastic giant cells (double arrow) (H and E, ×200). (b) Part of bony trabaculae, osteoid, epithelioid ostoblasts along with tripolar mitotic figure (single arrow) (H and E, ×400)

than on its microscopic features.[6] Therefore, while epithelioid osteoblasts are critical to the diagnosis of AO, tumor size and location seem to be more important considerations as far as treatment and prognosis are concerned. It should be remembered that tumors greater than 4 cm in diameter or those located in anatomic sites that impact the surgeon's ability to completely remove it are more likely to recur or cause local tissue destruction.[6,15]

Radiologically, our patient presented with an expansile lytic mass with internal calcifications and thin peripheral rim that appeared broken at places. The radiological findings in OB are thought to be non-specific.[13] They may have a central hyperdensity with surrounding halo just as OO. As these tumors are highly vascular, secondary aneurysmal bone cyst like changes may occur and cause diagnostic dilemma. However, unlike true ABC, there would be areas where the tumor would have entirely solid areas.[17]

From the treatment point of view, gross total excision is desirable in OBs in general. Complete excision can be achieved either by curettage or by en block resection, e.g., spondylectomy. Total excision reduces chances of recurrence and decreases the likelihood of malignant conversion. However, in patients like ours, complete excision is often not possible by dint of tumor location. Hence, adjuvant therapy like chemo/radiotherapy has a role to play in AOs. Role of chemotherapy is not as clear as that of radiotherapy. Adjuvant therapy is advocated in unresectable tumors, incomplete excisions, tumor with aggressive histologies and recurrent tumors.[18-20] Long-term relapse free survival of up to 25 years have been reported after radiotherapy.[21] Although, there is controversy as to what should be the optimal dose of radiotherapy (60 Gy vs. 40 Gy), a dose of 50 Gy given in a standard fractionated fashion (2 Gy, 5 days a week for 5 weeks) is probably the most effective regimen.[22] Radiotherapy usually causes either stabilization of tumor growth or its partial

reduction, rarely, if ever complete tumor resolution. Tumor response to radiotherapy immediately results in pain relief and causes ossification of the affected area in the longer run. There are, however, concerns with radiotherapy. These include chances of radiation induced bone necrosis with frank malignant degeneration in the residual tumor[12] and radiation myelitis in spinal AO's like in our patient. Chemotherapy is infrequently used in OBs in general. The indications are same as radiotherapy and people who advocate chemo, do so to avoid the concerns with radiotherapy mentioned above. Methotrexate appears to be an effective agent although polytherapy (methotrexate, doxorubicin and cisplatin) have also been used. Progression free survival up to 33 months have been reported with combination chemotherapy.[19,23]

In spite of all kinds of treatment discussed above, AO remains a difficult proposition with recurrences, malignant degeneration and finally an inevitable death. Although, recurrence rate in OBs in general is around 10% after treatment, it is as high as 50% with AOs.[6] Malignant degeneration eventually occurs with multiple recurrences and administration of radiotherapy.[6,12] This underscores the need for an active follow-up schedule for these patients and further research for other treatment options.

CONCLUSION

AO may affect craniovertebral junction and should be kept as differential diagnosis while dealing with extradural bony lesions in this location. We stress the need for meticulous histopathological examination in such cases for proper diagnosis and prognostication. Maximal surgical debulking followed by adjuvant chemo/radiotherapy appears to be the optimal treatment strategy as of now. These patients should be stringently followed-up as they have high chances of tumor recurrence/progression.

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