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Case Report

Basioccipital bone osteochondroma growing into the foramen magnum

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Received: 16 December 11

Accepted: 18 January 12

Published: 15 February 12

This article may be cited as:

Lotfinia I, Vahedi P, Tubbs RS, Gavame M, Vahedi A. Basioccipital bone osteochondroma growing into the foramen magnum. Surg Neurol Int 2012;3:21. Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2012/3/1/21/92937

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Abstract

Background: Osteochondroma is a common bone tumor and rarely affects the central nervous system. Although intraspinal osteochondromas are known to cause neurological deficits, intracranial osteochondromas with neurological compromise are very rare.

Case Description: The authors report an exceptional case of a quadriparetic 73-year-old patient with a basioccipital bone osteochondroma growing into the foramen magnum. The embryology, differential diagnoses, and optimal management strategies are discussed.

Conclusion: Although extremely rare, osteochondromas should be included in the differential diagnoses of tumors within the foramen magnum. For the tumors originating from the basioccipital bone, a simple medial suboccipital approach might suffice, while for ventral tumors, a far lateral transcondylar approach is necessary to avoid any neurovascular complications. Despite potentially catastrophic presenting symptoms, these tumors are pathologically benign and complete excision often results in long-term cure. To the best of our knowledge, this is the first report of an osteochondroma arising from the basiocciput.

Key Words: Foramen magnum, osteochondroma, skull base, suboccipital

Access this article online Website: www.surgicalneurologyint.com DOI: 10.4103/2152-7806.92937 Quick Response Code:

INTRODUCTION

Osteochondroma is a common benign tumor of bone. [11,22] The occurrence of the tumor as an intracranial mass is a very rare phenomenon. [1,3,8,9,18,23] The estimated incidence is only 0.1–0.2% of all intracranial tumors. [12] Such tumors show a predilection to the skull base, [9,21] probably due to the presence of multiple synchondroses here. However, unusual origins such as the convexity dura have also been

described. [8,21] Symptomatic tumors have been reported at the skull base, [1-3,23] dural convexity, [8,21] sella turcica, [8] occipital condyle, [16] clivus, [6] and cerebellopontine angle. [4] The tumor can also present as a craniofacial lesion. [10,25]

The occurrence of an intracranial osteochondroma is a rarity in the neurosurgical literature, [1,3,8,9,18,23] and to the best of our knowledge, this is the first description of a basioccipital bone osteochondroma growing into the foramen magnum.

CASE REPORT

History and physical examination

A 73-year-old male patient presented with gradual gait difficulty and four-limb weakness. The patient also complained of persistent dull pain in the occipitocervical region. There was no history of previous trauma. The patient was under treatment for hypertension and had a history of lumbar laminectomy and discectomy several years ago. Physical examination revealed spastic quadriparesis (3/5 on the right and 2/5 on the left), positive Hoffmann's sign, extensor plantar response, and increased deep tendon reflexes. The gait appeared to be spastic, but cerebellar examinations were within normal range. Although the patient was complaining of tingling in all extremities, no apparent deficit was found during routine sensory examinations.

Imaging

With a high suspicion of a space-occupying lesion, brain magnetic resonance imaging (MRI) was performed, which revealed a tumor with bony characteristics at the level of the foramen magnum. The tumor created significant compression of the cervicomedullary junction [Figure 1a]. 3D bone computed tomography (CT) scan confirmed the lesion to arise from right suboccipital bone containing both cortical and cancellous elements [Figure 1b and c]. On CT myelography, cord compression was apparent [Figure 1d].

Surgery

The patient underwent a right medial suboccipital craniectomy under general anesthesia in the prone

position. A Mayfield clamp was applied and the midline incision was made from just above the inion to the C2 spinous process. The suboccipital muscles were dissected subperiosteally on the right side. A small right medial suboccipital craniectomy plus unilateral removal of the C1 posterior arch was done to visualize all parts of the tumor, which necessitated removing the posterolateral rim of the foramen magnum. The solid bony lesion was resected totally using air drill and rongeur. Fortunately, the tumor had no encroachment on the dura mater of the posterior fossa or right vertebral artery and was completely resected.

The postoperative course was uneventful and the patient had significant recovery immediately after the operation. Postoperative CT scan confirmed removal of the lesion [Figure 1e].

Pathology

The pathological examination of the specimens revealed mature bone trabecules covered by the cartilaginous tissue with no atypia or mitotic figure, in favor of an osteochondroma [Figure 1f].

Follow-up

At 18 months follow-up, all of the patient's symptoms resolved except for intermittent headache, which might be attributed to the craniectomy site. Mild limb spasticity remained.

DISCUSSION

Osteochondroma is the most common tumor of the

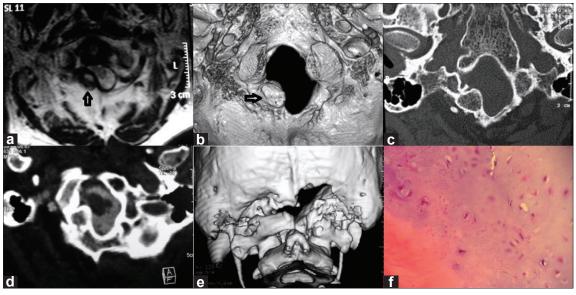


Figure 1:(a)TI-weighted axial MRI of the craniocervical junction at the level of the foramen magnum shows considerable cord compression due to a lesion (arrow) on the right side of the occipital bone. (b) 3D reconstructed CT scan at the same level confirms the bony nature of the lesion (arrow). (c) The tumor contains both membranous and cortical bone. (d) CT myelography at the level of the foramen magnum also shows cord impingement by the tumor. (e) Postoperative CT scan depicting the extent of resection. (f) Pathology confirmed the lesion as an osteochondroma

bone, which constitutes 10–15% of all bony tumors and 20–50% of benign bone tumors. [10,15] The lesion is an exophytic bony protrusion covered by a cartilaginous cap, [11,22] which is most commonly found in long bones, and especially at the epiphysis. Nearly 40% of cases are seen around the knee joint. [22] The location of the tumor within the central nervous system is not common. The spinal column is the predominant location for such tumors when they involve the central nervous system [14,15] and these tumors around the brain are quite rare. [1-3,8,9,18,23]

When these tumors are found at the skull base, it is thought that they arise from remnants of the cartilaginous parts of the basilar primordial synchondrosis that are trapped during enchondral ossification of the skull base. This might explain why these lesions tend to appear within the middle cranial fossa, especially near the basioccipital and basisphenoid synchondroses. [8,12,17] Also, it might explain the predominant extracranial location of the tumor; however, in extremely rare cases, these tumors might arise from the dura mater of the convexity or falx cerebri. [8,21] Our case demonstrates the rare presentation of an intracranial osteochondroma.

As described by Shapiro and Robinson, [24] the occipital bone in humans originates from four distinct cartilaginous centers, except for the interparietal region which has a membranous origin. The four cartilaginous centers are located at the basioccipital, exoccipital (two centers), and supraoccipital areas. The ossification of these centers is started during the first trimester of pregnancy and continues into early childhood. As for the present case, the possible origin of the tumor should be from the cartilaginous remnants of the supraoccipital center. The different embryology of the intraparietal basiocciput explains why the tumor was confined only to the skull base portion of the occipital bone. Nevertheless, Lichtenstein^[13] believed that osteochondromas might originate from any kind of bone because of the pluripotential nature of the periosteum, which enables it to produce both the bony and cartilaginous tissues.

Usually, skull osteochondromas are solitary; however, multiple skull exostoses have been described in Proteus syndrome. This syndrome presents with mental retardation, multiple central nervous system anomalies, hemimegalencephaly, macrodactyly, osteochondromas, and soft tissue tumors. Likewise, multiple lesions might be seen in association with other mesenchymal tumors like Maffucci and Ollier syndromes. [8,12]

Osteochondroma might become symptomatic due to the mechanical irritation of cranial nerves, soft tissues, or vascular compression, injury, or fracture. [8,9] As in our case, the presence of the tumor within the foramen magnum might be immediately noted due to the occipital headache and compromised neurovascular structures in this area. In incidental and asymptomatic patients,

observation might suffice; [14,15] but for symptomatic patients, surgery is warranted because these lesions are resistant to chemoradiotherapy. [3]

Other bony lesions that should be considered in the differential diagnoses include intraosseous meningioma, monostotic fibrous dysplasia, osteoma, osteoblastoma, osteoblastic metastases, giant cell tumor, and eosinophilic granuloma. Although rare, it is imperative to differentiate the tumor from unilateral atlantal lateral mass hypertrophy before surgery, as atlantoaxial fixation might be necessary after the decompressive procedure. Careful preoperative review of reconstructed CT of the craniocervical region makes this distinction possible.

CONCLUSION

Although extremely rare, osteochondromas should be included in the differential diagnoses of tumors within the foramen magnum. Depending on its origin from the remnants of the different occipital cartilaginous centers around the foramen magnum, such tumors might present as ventral, lateral, or dorsal masses. For the tumors originating from the basioccipital bone, a simple medial suboccipital approach might suffice, while for ventral tumors, a far lateral transcondylar approach is necessary to avoid any neurovascular complications. Despite potentially catastrophic presenting symptoms, these tumors are pathologically benign and complete excision often results in long-term cure.

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