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GIANT OSTEOCHONDROMA OF THE ANTERIOR CRANIAL FOSSA - A RARE ENTITY

EMMANUEL THAS J
Department of Neuro Surgery,MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : BackgroundOsteochondroma 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 DescriptionThe authors report a case of 21 year old patient with a base of the anterior cranial fossa osteochondroma causing neurovascular compromise. The embryology, differential diagnoses, and optimal management strategies are discussed.ConclusionAlthough extremely rare, osteochondromas should be included in the differential diagnoses of tumors within the skull base. Despite potentially catastrophic presenting symptoms, these tumors are pathologically benign and complete excision often results in longterm cure.

Keyword : Osteochondroma, skull base, anterior cranial fossa

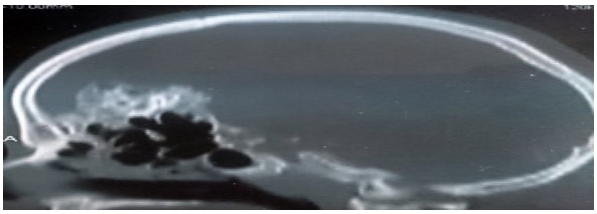
INTRODUCTION

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] more commonly in the middle cranial fossa, probably due to the presence of multiple synchondroses here. However, unusual origins such as the convexity dura, air sinuses 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.[10,24] We present a case of osteochondroma of the base of the anterior cranial fossa in a 21 year old male and discuss the current literature.

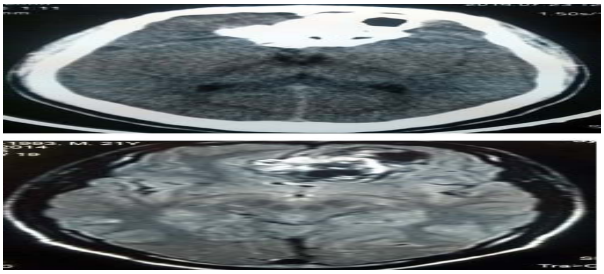
CASE REPORT

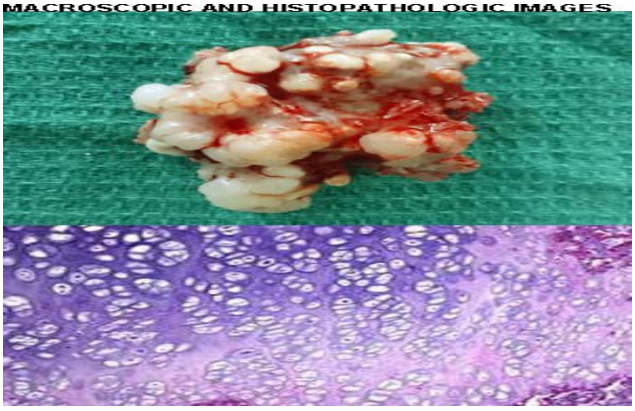
A 21 year old male with no known comorbidities came with complaints of headache for past 1 year, anosmia of both the nostrils for past 6 months , 2 episodes of generalised tonic clonic seizures in a period of 1 month with an interval of 15 days. The patient did not have any other symptoms of visual disturbances, facial numbness or facial asymmetry, with no symptoms of lower cranial nerve palsy or weakness of upper limb or lower limbs or numbness.

The patient on examination showed MMSE of 28/30 and was not able to appreciate smell in both the nostrils with intact other cranial nerve functions and spinomotor system and sensation. the patient did not have any cerebellar signs. patient had underwent CT brain plain and contrast which showed a hyperdense lesion in the floor of the anterior cranial fossa compressing on the frontal lobe and found to be arising from the spheno-ethmoidal sinus. MRI brain also showed a T1 and T2 heterointense lesion in the anterior cranial fossa involving the frontal , ethmoidal and sphenoidal sinus. a provisional diagnosis of bony lesion of the skull base was done and planned for total excision of the lesion . The patient underwent bifrontal craniotomy with total excision of the lesion which was found to be arising from the sphenoethmoidal sinus pearly white in colour with mulberry appearance . Histopathology of the specimen came to be osteochondroma of the skull base

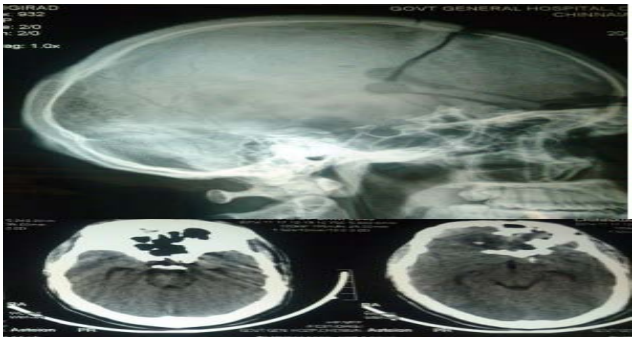


PRE-OP XRAY, CT AND MRI BRAIN





POST OP IMAGES



DISCUSSION

Osteochondroma is the most common tumor of the 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. 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.[12] This syndrome presents with mental retardation, multiple central nervous system anomalies, hemimegalencephaly, macrodactyly, osteochondromas, and soft tissue tumors.[5] 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 in the base of the anterior cranial fossa caused 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,[19] osteoblastoma,[20] osteoblastic metastases, giant cell tumor, and eosinophilic granuloma.

CONCLUSION

Although extremely rare, osteochondromas should be included in the differential diagnoses of tumors in the base of the anterior cranial fossa. Depending on its origin from the remnants of the different cartilaginous centers around the base of the anterior cranial fossa foramen magnum, such tumors might present with varied neurovascular compression syndromes. Despite potentially catastrophic presenting symptoms, these tumors are pathologically benign and complete excision often results in longterm cure.

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