Osteochondroma of Clivus; An Unusual Cause of Headache
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Introduction

Osteochondromas are benign bone tumors of mesenchymal and non mesenchymal types (1). Their presentation as intracranial mass is a very rare phenomenon (2). The incidence is only 0.1-0.2% of all intracranial tumors. Hirschfield first reported intracranial chondromas in 1851 (3). But first well documented, case was reported by Levitt in 1934 (4). Predilection to the base of skull is noted possibly because of the presence of multiple synchondroses. They were reported in different areas including sella turcica, occipital condyle, Cerebellopontine angle and dural convexity. Craniofacial lesions were also reported. We present a case of significantly large osteochondroma of clivus presenting as trivial finding like headache, requiring surgical removal of the tumor with resolution of symptoms. Only one case of osteochondroma in the clivus was reported before this case, to the best of our knowledge (5).

Case Report

It showed osseous pedunculated lesion with cartilage cap making it radiologically and characteristically typical osteochondroma on CT scan. There was a bony projection arising from the posterior wall of the clivus impinging into the pons tissue. The cerebral hemispheres and cerebellum appeared normal with normal gray-white matter differentiation without any signs of brain edema or any focal lesion. The clival lesion appeared to be of primary bony etiology and due to presence of heterogenous calcified matrix a diagnosis osteochondroma was considered with differential diagnosis of chordoma and chondrosarcoma.

MRI of brain (figure 2) was carried out for further assessment of the lesion and specially to evaluate the pontine tissue impingement. On MRI the lesion was showing predominantly hypo-intense signal on both T1 and T2 weighted images, and hyper-intense signal on T2 weighted images. On post contrast images there was no significant enhancement of lesion.

Figure 1: CT Scans showing clivus bony projection (arrow)
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Case Report

The lesion was causing slight lateral displacement of basilar artery without any encasement or compression. The bony spur like projection from the posterior part of the lesion was projecting into the left side of pons for a depth of 9 mm but there was no edema or mass effect within pontine parenchyma. There was no extension into the sphenoid sinus and sella turcica and the pituitary gland was normal. No associated soft tissue mass was seen with the clival lesion.

Histopathological examination (figure 3) showed a fragmented cellular osteocartilagenous lesion. There were fragments of cellular cartilage with intermingled bone trabeculae having marrow spaces. The intact zonation was seen in case due to fragmented nature of the specimen but the cellular cartilage with bone and marrow, all three components were present. Histological picture with the radiological correlation was consistent with osteochondroma.

Figure 2: MRI brain, (A, B) Axial T2 and T1 weighted images and (C) midline sagittal T1 weighted image, showing a mass in clivus

Later trans-sphenoidal excision and biopsy was done from the lesion. Dural defects were sealed with duro-seal, facial and fat graft harvested from left thigh. Post operatively she was kept on acetazolamide, labyrinthine sedatives, analgesics and antibiotics. She made uneventful recovery with complete resolution of her headache and was discharged three days after the procedure.
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Discussion
An osteochondroma is a cartilage-covered bony excrescence (exostosis) that arises from the surface of a bone. They account for 10-15% of all bony tumors and 20-50% benign bony tumors (6). Long bones, especially epiphysis, are the most common site of these tumors. These tumors are very rare in skull (2). In central nervous system, spinal cord is the main site of occurrence (6). Lesion is characterized by a cartilaginous cap over exophytic bony protrusion (7). Remnants of trapped cartilaginous parts of basilar primordial synchondrosis during enchondral ossification of basal skull, give rise to these tumors at the base of skull. This explains usual position of these tumors near basi-occipital and basi-sphenoid synchondrosis in the middle cranial fossa (8). Our case represents a rare location of intracranial osteochondroma. According to Lichtenstein, any type of bone can give rise to osteochondroma because of the pluripotential nature of peristeam leading to the development of both cartilaginous and bony tissues (7). Different embryological origin of intra-parietal basi-occiput explains the reason of confinement of these lesions to the occipital bone of basal skull (2). Osteochondromas can be multiple as in Proteus Syndrome, but usually solitary. Maffucci and Ollier syndromes have multiple lesions in association with other mesenchymal tumors (2). Mechanical irritation of soft tissues, cranial nerves, vascular compression or fracture can lead to symptomatic presentation of osteochondroma (8). Asymptomatic incidental lesions can simply be observed (6). Monostotic fibrous dysplasia, intra-osseous meningioma, osteoma, osteoblastoma, giant cell tumor and eosinophilic granuloma are the differentials. They occur as either solitary intracranial tumor or in combination with skeleton chondromatosis.

Commonly occurs in females in 2nd and 5th decades of life. Clinical signs are usually due to cranial nerves involvement. They are very slow growing tumors, histologically benign but have serious implications due to close proximity of vital intracranial structures (5). Even sub-total surgical resection provides great benefit due to close proximity of vital structures at the base of skull. They are radiotherapy resistant. The site of the tumor determines prognosis (5). During development, skull vault develops from membranes but bones of skull base arises from cartilaginous tissues which explains increase frequency of cartilaginous tumors at the base of skull arising from cartilaginous rests either between centers of ossification or in the suture lines (9). Four main groups of these tumors are those arising from sinuses, meninges, base of skull, or in brain substance (10). Conventional radiography is the study of first choice for radiological investigation. In plain X-ray these lesions may appear as granular calcifications, sometimes as sclerotic areas resembling cancellous bone. Calcification along with adjacent local bone erosion is typical of osteochondroma. CT can be used to determine if the marrow and cortices of the lesion are continuous with the parent bone. The role of MRI is predominantly, to assess surrounding soft tissues and to measure the thickness of the cartilage cap, which can be important in evaluating for malignant generation. Less than 1% of solitary osteochondromas undergo malignant degeneration of the cartilage cap into secondary chondrosarcoma. The risk of malignant degeneration increases with an increase in the number and size of the osteochondromas. In general, a sessile lesion is more likely to degenerate into sarcoma than a pedunculated lesion.
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Conclusion

Although histologically benign and extremely rare osteochondromas can present as trivial findings like migraine type of headache. Relevant radiological investigations should be done in emergency department to rule out such causes. Surgery is required for symptomatic patients due to close proximity of vital intracranial structures and due to their radio and chemotherapy resistance. Even sub-total resection provides great benefit. Asymptomatic incidental lesions can simply be observed.

References