Osteochondroma of basi-occiput

Sir,

Osteochondromas are common ‘benign’ tumors involving the long bones.[1-4] Some authors consider it as a developmental bony lesion rather than a true neoplasm.[1-4] Osteochondromas rarely affect skull bones[5] and only anecdotal reports are available in the literature.[4-6] Management issues for cranial lesions are unclear. The case is discussed and the literature on the subject is briefly reviewed.

A 20-year-old male presented with eight months history of progressive weakness, stiffness and tingling sensations over right half of the body and hoarseness of voice. There were no cutaneous stigmata or any significant family history suggesting a congenital lesion. He also had a painless, bony hard swelling involving the head of the right tibia, for about a year. Clinical examination revealed depressed gag and pharyngeal reflexes on the right side and spastic quadriparesis, right side being worse. Investigations revealed a large predominantly bony lesion located in the anterior basiocciput causing deep indentation and deformity of the brainstem and the cerebellum [Figures 1-3]. The jugular bulb region was involved. The extradural lesion was exposed by a retro-mastoid route. The entire tumor was firm and crystal like bone. Tidious drilling and bone cutting instruments were used extensively with considerable difficulty. The proximity to the region of the jugular bulb, internal auditory meatus and to the brainstem made the surgery a formidable endeavor. The lower cranial nerves could not be identified within the tumor. A partial resection of the tumor was done sufficient to decompress the region of the brainstem and cerebellum. Postoperatively, the patient developed significantly enhanced symptoms of ipsilateral lower cranial nerve paresis. During the postoperative

Figure 1: Axial CT scan (bone window) image showing a large bony excrescence arising from the basiocciput

Figure 2: Sagittal T-1 weighted image showing the lesion arising from the basiocciput and extending into the posterior fossa

Figure 3: Axial T-2 weighted MR image showing an extra-axial lesion indenting into the brainstem and cerebellum. The lesion is hyperintense towards the tip – a feature indicative of the cartilaginous cap
phase, he needed tracheostomy and subsequently developed pulmonary infection. He died on the twelfth postoperative day. Histology revealed characteristic features of osteochondroma. The osteocartilaginous tissue had a cartilage cap that merged with the underlying bone [Figure 4].

Osteochondromas are amongst the more common benign tumors of the bone.[2] They account for approximately 20-50% of all the benign tumors and about 10-15% of all the bone tumors.[1,3] Several authors have suggested that these tumors probably represent a developmental lesion rather than a true neoplasm.[1,3] Osteochondromas are composed of cortical and medullary bone with an overlying hyaline cartilage cap and demonstrate continuity with the underlying parent bone cortex and medullary canal.[1-3] Osteochondromas may be solitary or multiple; the latter being associated with the autosomal dominant syndrome called hereditary multiple exostoses (HME). Apart from cosmetic deformity, rarely, features like fracture, vascular compromise, neurologic sequelae, overlying bursa formation and malignant transformation are observed.[2] Malignant transformation is seen in approximately 1% of solitary osteochondromas and in 3-5% of patients with HME.[2] Hyaline cartilage cap thickness is an important criterion in determining malignant transformation.[5] Hereditary multiple exostoses, also known as familial osteochondromatosis or diaphyseal aclasis, is characterized by the development of multiple osteochondromas. Approximately two-thirds of affected individuals have a positive family history. Genetic abnormalities have been identified, with three distinct loci: one each on Chromosomes 8, 11 and 19.[3]

Cranial osteochondromas are extremely rare. We could isolate two such cases from the literature, one located in the atlantoaxial joint and the other in the region of the falx cerebri.[4,6] Surgical resection is the only form of treatment for such lesions as they are resistant to treatment by radiotherapy and chemotherapy.[3] Although radical total resection of the lesion is reported to be curative in nature, it is unclear if such a treatment was possible in the presented case. Surgery in the presented case was contemplated due to the progressive nature of symptoms and due to the symptomatic involvement of the lower cranial nerves. During surgery we observed that the tumor was much more firm and elastic in nature than the normal bone, making both drilling and cutting extremely difficult and tedious. The proximity of the lesion to critical regions of the brain made the surgery a formidable technical problem.

Although rare, osteochondroma can affect the skull base. Due to the benign nature and possibility of cure, it is tempting for a surgeon to attempt radical resection. Hard and elastic consistency of the tumor must be borne in mind prior to contemplating surgery.

Vivek Bonde, Balasubramaniam Srikant, Atul Goel
Department of Neurosurgery, Seth G.S. Medical College and King Edward VII Memorial Hospital, Parel, Mumbai - 400012, India

References


Accepted on 16-09-2006