Intradiploic cavernous hemangioma

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Cavernous hemangiomas of the skull are uncommon bone tumors with an incidence of 0.7-1% with majority occurring in the calvaria and vertebrae. Frontal bone and parietal bones are common location in the skull with a female preponderance. Occipital bone location is very rarely reported with these benign bony vascular lesions [1)].

They are supplied by the branches of the external carotid artery in the diploic space. Intradiploic hemangiomas usually erode the outer table of the skull and despite tumour enlargement, inner table remains intact [2)].

A primary intraosseous hemangioma (IOH) of the orbital bone is extremely rare. The preferred method of treatment for IOH is total surgical excision with reconstruction. Herein, the authors describe a patient with an orbital roof IOH and the unexpected complications of ptosis and deteriorated exophthalmos. These findings showed that the total surgical excision and subsequent reconstruction provided adequate decompression and prevented further ocular complications from the orbital wall defect [3)]

Etiology

Etiology is believed to be congenital origin or previous trauma. These arise from vessels present in the intradiploic space and derive their blood supply from the branches of external carotid system such as middle meningeal artery and superficial temporal artery [4)].

Clinical features

These tumors are slow growing and are generally asymptomatic [5)].

The major clinical problem associated with these tumors is blood steal from brain parenchyma and resultant atrophy and mental retardation. Other may be congestive heart failure and thrombocytopenia known in this setting as Kasabach–Merritt syndrome [6) 7) 8) 9)].

Diagnosis

Histopathologic confirmation of the tumor is the definitive method for diagnosis.

Plain Radiographs

On plain radiographs, they show sunburst of radiating trabeculae having peripheral sclerotic rim. This is due to gradual erosion of surrounding bone as they grow.

CT

It is a lytic, well-defined defect characterized by a fine, reticular pattern without a sclerotic rim, involving both inner and outer tables on CT scans.

MRI

The MRI signal is variable depending on the amount of fat content.

On MRI, these lesions are hyperintense on T1- and T2-weighted imaging (WI) with intense post-contrast enhancement. However, the intensity on T1-WI is variable depending upon the presence of fat.

The hyperintensity of hemangiomas at T1-weighted sequences is an important distinguishing feature for this tumor [10].

Can be seen as as multiple craters on skull [11]

Differential diagnosis

Although they are benign, radiological findings are not always characteristic and their multiple presentation may easily make surgeons consider the other malignancies of the skull in the differential diagnosis.

The common differentials include aneurysmal bone cyst, giant cell tumor, Langerhans' cell histiocytosis, sarcoma, meningioma, metastatic disease, and dermoid tumor [12].

Treatment

Complete surgical excision is the treatment of choice. Most important goal at the time of surgery is control of bleeding. Preoperative embolization is one option. Other strategy is excision of the tumor with surrounding rim of normal bone without entering into the tumor [13].

Case reports

2014

A 15-year-old male, who had prominent inion since early childhood, presented with fall on occiput followed by transient loss of consciousness to our emergency department. Neurological examination was normal, except a nontender unduely prominent inion. Axial computed tomography (CT) revealed a focal bony swelling at the external occipital protuberance, without any parenchymal abnormality.

Biopsy of the lesion was advised, but patient and relatives ignored the medical advice. Patient reported to us after 18 months when he developed severe occipital headache on lying supine. The pain forced him to sleep in lateral postures only. Local examination revealed tenderness and significant increase in size of swelling at the external occipital protuberance measuring 7 × 6 × 6 cm in size. Neurological examination was normal. CT brain showed an expansile lytic lesion involving the occipital bone having a coarse bony matrix and radiating spicules. There was erosion of both inner and outer tables of skull. Magnetic resonance imaging (MRI) revealed a T2 heterogeneously hyperintense, T1 isointense, and intensely enhancing mass with intracranial extradural extension. A large soft tissue component in the scalp was also present. Surgical excision of the lesion was done with margin of surrounding normal bone. The lesion was intradiploic in location with expansion of outer and inner tables of skull. The underlying dura was intact, however, mass effect on venous sinuses was present. Careful preservation of the torcula and venous sinuses was done. The bone defect was filled with appropriately tailored bone cement. Histopathology revealed anastomosing, mainly thin-walled vascular channels lined by a single layer of flattened to plump endothelial cells. The tumor cells were seen amidst the bony trabeculae. These findings were suggestive of intraosseous cavernous hemangioma. Postoperatively, patient was relieved of his pain and was able to lie supine comfortably. Follow-up CT of head showed complete excision of the lesion without recurrence [14].

2012

A 27-year-old man presented with a large hard left frontal swelling. Computed tomography (CT) of skull showed a large, expansile lesion destroying both inner and outer tables of the skull. The lesion had a larger intracranial component with mass effect and showing homogenous, intense enhancement with contrast. Magnetic resonance imaging (MRI), in addition to the above features, showed the lesion to be purely intradiploic having both extra- and intracranial extension. The adjoining dura showed a characteristic enhancement, dural tail sign. The bone window showed gross calcification of the lesion. The pre-operative diagnosis was meningioma. On MR venography, the mass was indenting the superior sagittal sinus however, the patency was intact. Intra-operatively the finding was of a large expansile, vascular, hard and gritty lesion with dural attachment suggestive of a primary calvarial meningioma. Histologically the lesion showed features cavernous hemangioma [15].

2011

A 31-year-old patient who had had frontal cephalalgias for several years. CT and MRI anatomical imaging objectified a frontal osteolytic tumor respecting the osseous external table but compressing the superior sagittal sinus. Total en bloc resection of the tumor associated with titan cranioplasty was performed. The postoperative course was uneventful. Three months after surgery the patient no longer reported headache. The anatomical and pathological results concluded in intradiploic cavernous hemangioma. We discuss this case and others described in the literature [16].

2008

A 16-year-old child who was admitted with a swelling lesion in the right parietal bone and diagnosed as cavernous hemangioma after total extirpation [17].

2004

A 50-year-old man in whom a symptomatic subdural hematoma (SDH) resulting from a cavernous hemangioma of the calvaria had hemorrhaged and eroded through the inner table of the skull and dura mater. The patient underwent surgery for evacuation of the SDH and resection of the calvarial lesion. Postoperatively, the patient experienced immediate relief of his symptoms and had no clinical or radiological recurrence. Calvarial cavernous hemangiomas should be considered in the differential diagnosis of nontraumatic SDHs. Additionally, skull lesions that present with intracranial hemorrhages must be identified and resected at the time of hematoma evacuation to prevent recurrences [18].

2003

A 42-year-old female patient with multifocal cavernous hemangioma of the skull associated with nasal osteoma. X-rays, computerized tomography, magnetic resonance imaging, and histopathology were used to achieve the diagnosis of this rare entity. The multiple cavernous hemangiomas were resected en-bloc and a curettage biopsy was obtained from the nasal osteoma. The patient healed well after the operation. No recurrences of the cavernous hemangiomas were observed after one-year follow-up. [19].

2000

A 3-year-old right hand dominant male presented with a non-tender parietal scalp swelling of a 1-year duration. History included a skull fracture located in the same region 24 months before presentation. Neurological examination was unremarkable. Pathological examination after curettage of the lesion revealed features consistent with organizing hematoma [20].

1993

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