

Secondary Aneurysmal Bone Cyst Developing In Fibrous Dysplasia

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Key words : Fibrous dysplasia, Secondary aneurysmal bone cyst.

Introduction:-

An aneurysmal bone cyst (ABC) is not a true neoplasm and may be a benign reactive vascular lesion. The exact etiology of this tumor is unknown but the descriptive name is derived from the microscopic appearance of a blood filled, expansile, sponge like tumor containing numerous giant cells.

Aneurysmal bone cysts are known to arise in association with other abnormalities of the skeleton, particularly non-ossifying fibroma, fibrous dysplasia and chondromyxoid fibroma. Such lesions have been described as secondary ABCs [1]. They have also been recorded following fractures [1]. The ABCs most commonly involve the metaphyses of long bones or the posterior elements of vertebrae, but the nasal cavity is the extremely rare site [2].

We report an interesting and rare case of secondary ABC developing in fibrous dysplasia in the nasal cavity & nasopharynx.

Material and method:-

A 13 yrs male child presented with enlarging right nasal mass. Radiograph of nasopharynx lateral view showed a nasal mass also extending posteriorly to obliterate the nasopharyngeal air column.

On CT scan (coronal) was seen a large (5.5 X 6cm) mass lesion attached to the base of anterior cranial fossa protruding into nasopharynx, right orbit and right nasal cavity (Fig-1) with deviation of the nasal septum to left side. The lesion had a very thick osseous rind (HU+350) & large cystic centre (HU+6 to +40) (Fig-1,2). There was mild sclerosis & marked expansion of body & right sphenoid wings from which the mass was presumably arising. An extension of the mass into right frontal sinus was seen. The mass was causing proptosis of right eyeball

and pressure moulding with sclerosis of medial wall of both orbits, both maxillary sinuses & ethmoid air cells suggesting slow growth & benignity of the lesion (Fig-1).

The roof of right orbit was elevated & a breach in the base of anterior cranial fossa on right was seen.



Fig 1

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Fig 2

Taking into account the typical location, thick osseous rind, expansile mixed lytic and sclerotic nature of the mass diagnosis of fibrous dysplasia was considered. The presence of a very large essentially cystic centre (Fig-1) helped us to reach to a diagnosis of fibrous dysplasia degenerating into an aneurysmal bone cyst.

The radiological findings were confirmed on excision biopsy of the lesion.

Discussion:-

Fibrous dysplasia is a disease of unknown etiology characterised pathologically by replacement of the medullary bone by well defined areas of fibrous tissue and cysts containing blood or serous fluid. The fibrous tissue then undergoes varying degrees of abnormal ossification so that some of the lesions show an increase in density, dependent on extent of ossification. This increase in density may thus be patchy, giving a cottonwool appearance, or homogenous giving a ground glass appearance [3].

Skull lesions often show asymmetric thickening of the vault with sclerosis at the base, with multiple areas of radiolucencies. A grossly expanded hyperostosis of sphenoid may resemble a meningioma. There may be expansion with obliteration of paranasal sinuses or disruption of teeth. Orbital fissures may be encroached upon & proptosis may result.

Etiology of ABC: The true etiology is unknown. Most believe that ABCs are the result of a vascular malformation within the bone; however, the ultimate cause of the malformation is a topic of controversy. Three commonly proposed theories are as follows [1]:

" ABCs may be caused by a reaction secondary to another bony lesion. This theory has been proposed because of the high incidence of accompanying tumors in 23-32% of ABCs. Giant cell tumors are most commonly present. However, many other benign and malignant tumors are found: fibrous dysplasia, osteoblastoma, chondromyxoid fibroma, nonossifying fibroma, chondroblastoma, osteosarcoma, chondrosarcoma, unicameral or solitary bone cyst, hemangioendothelioma, and metastatic carcinoma. ABCs in the presence of another lesion are called secondary ABCs. ABCs may arise de novo. ABCs with no evidence of another lesion are classified as primary ABCs.

" ABCs may arise in an area of prior trauma.

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