

An Aneurysmal Bone Cyst Arising From Fibrous Dysplasia In Frontal Bone

RD DANI, VS GANDHI, GN THAKKAR, HJ PRAJAPATI, GD RATHOD

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Introduction: -

Fibrous dysplasia of the cranium is a relatively uncommon condition that affects primarily the anterior cranial region; its occurrence in the cranial base in combination with aneurysmal bone cyst (ABC) constitutes an extremely rare condition. [1]

Here, we report a case of fibrous dysplasia involving paranasal sinuses, skull base, floor of right side of anterior cranial fossa and right side of frontal bone with secondary aneurysmal bone cyst arising from frontal bone on right side. Only two such cases have been reported previously in the literature.



Figure 1: - Photograph of patient's face showing swelling on right side of head and prominent right supra orbital ridge and hypertelorism.

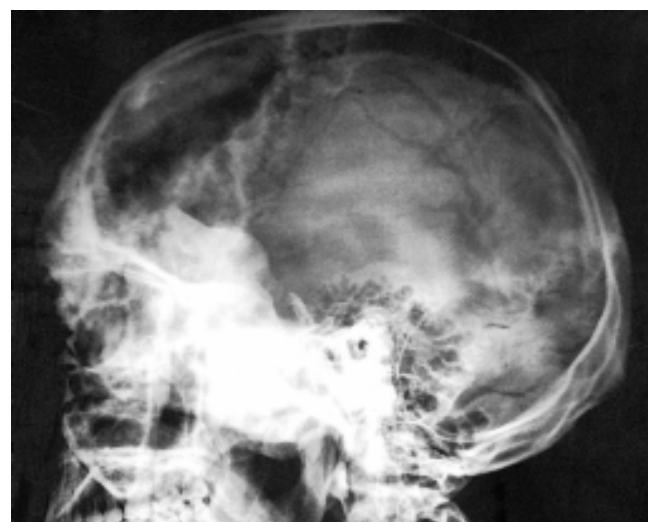


Figure 2, 3: -Radiograph of skull in AP and lateral view revealed expansion, sclerosis and thickening of base of skull, frontal bone, and ethmoid bone, mainly on right side. Osteolytic lesion with soft tissue swelling and thinning and destruction of outer table of skull vault was seen on right fronto-parietal region.

From the Department of Radiology & Imaging, Smt. S.C.L. Hospital Saraspur, Smt. N.H.L Municipal medical college, Ahmedabad(Gujarat)- 380 006, INDIA

Request for Reprints: Dr.Ranna D Dani, Professor & Head, Radiology Department*, 83, Yogeshwarnagar Society, Opp, Anjali Theater, Near Dharnidhar Society, Bhatta, Paldi, Ahmedabad-380007 Gujarat, INDIA.

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Case Report: -

A 23 year old male patient came to hospital with chief complain of sudden swelling on the right side of skull with headache since last 15 days.

On clinical examination swelling was cystic in nature. The patient had facial deformity since several years, having prominent right supraorbital ridge and hypertelorism looked like a 'leonine face'. {Leontiasis ossea} (Figure 1)

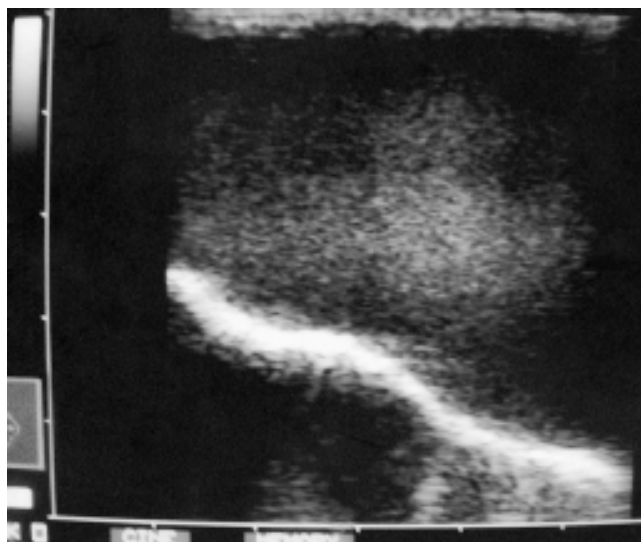


Figure 4: - USG of the swelling is showing fluid with marked internal echoes.



Figure 5: - CT Scan of skull (bone window) showing sclerotic frontal bone on right side with focal expansile cystic lesion noted in it

Radiograph of skull in anteroposterior and lateral view revealed expansion, sclerosis and thickening of base of skull, frontal bone, and ethmoid bone, mainly on right side. Osteolytic lesion with soft tissue swelling and thinning and destruction of outer table of skull vault was seen on right fronto-parietal region. (Figure 2, 3)

USG of the swelling was performed with five MHz linear probe, and showed moving fluid with marked internal echoes. (Figure 4) Then, aspiration of the fluid revealed blood. ACT Scan of skull with facial bones was performed. It showed expansile spongiform sclerotic bony lesion involving ethmoid, sphenoid and frontal bones. There was focal expansile cystic lesion noted at frontal bone on right side with fluid level noted in the cyst, with localized thinning of inner table of skull vault. There was relative sparing of temporal and parietal bones. (Figure 5, 6)

The patient was operated, excision of the swelling was done and the biopsy was taken from the margin of the lytic area. Histopathological examination showed changes of aneurysmal bone cyst associated with fibrous dysplasia. (Figure 7)



Figure 6: - CT Scan of skull showing cystic lesion in frontal bone on right side with fluid level noted in the cyst.

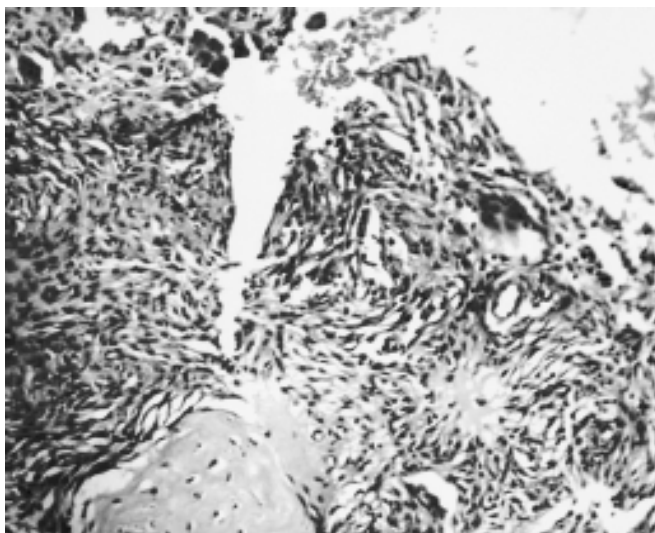


Figure 7: - Microscopic appearance of lesion showing blood filled space with areas of haemorrhage and multiple osteoclast like giant cell. Bone tissue without osteoblastic rimming also seen.

Discussion: -

An aneurysmal bone cyst is neither an aneurysm nor a true cyst but it does occur in bone. Rather, it is a benign, non-neoplastic, expansile osseous lesion. [2] The exact etiology of this tumor is unknown but the descriptive name is derived from the macroscopic appearance of blood filled expansile sponge-like tumor containing numerous giant cells. Aneurysmal bone cyst has been shown to arise in association with other abnormalities of the skeleton, particularly non-ossifying fibroma, fibrous dysplasia and chondromyxoid fibroma. Such lesion has been described as 'secondary' aneurysmal bone cysts. [3].

It can present as a slowly or rapidly enlarging mass with non-throbbing pain. Between three to twelve percent of aneurysmal bone cysts occur in the head and neck, and they also have been reported in the maxilla, orbit, ethmoid and frontal bones. [2]

Fibrous dysplasia is a skeletal developmental anomaly of bone forming mesenchyme in which osteoblasts fail to undergo normal morphological differentiation and maturation. It is of unknown cause and not hereditary and it may affect one bone, a few bones or many bones. Involvement of the skull and lesions of the facial bones occur with nearly equal frequency and are noted in approximately 10 to 25 % of patients with monostotic fibrous dysplasia and in 50 % of those with polyostotic involvement. [4] In our case the patient is of monostotic variety.

Bonakdarpour A et al analyzed radiological findings in 75 cases of aneurysmal bone cysts. In them, 65 % were primary and 35 % were secondary, the aneurysmal bone cyst being combined with other osseous lesions. [5] Martinez et al reviewed a group of 639 bone lesions in order to study the features of aneurysmal bone cysts and its association with other conditions. They found primary aneurysmal bone cyst in 87 patients and secondary aneurysmal bone cyst in one patient out of 42 patients of fibrous dysplasia. [6]

The co-existence of these two lesions supports the theory that aneurysmal bone cyst may represent secondary change due to haemodynamic alteration of the vascular bed caused by fibrous dysplasia. [7]

Conclusion: -

The diagnosis of aneurysmal bone cyst should be considered in the patient with fibrous dysplasia that has a cystic component or in patients of fibrous dysplasia who present with sudden expansion of the lesion. [8]

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