Fibrous Dysplasia of Ethmoid

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Fibrous dysplasia is a peculiar clinical entity, which is of interest to the Otolaryngologist, because at times it involves one of the paranasal sinuses.

Fibrous dysplasia of the bone is the name applied by Lichtenstein (1938) to a polyostotic or monostotic condition apparently due to a perverted activity of the specific bone-forming mesenchyme. It is most frequently localized in the femur, tibia, rib, maxilla and mandible. Among the extra-skeletal aspects of the condition, abnormal pigmentation of the skin, premature sexual development in females and signs of hyperthyroidism have been described in very severe conditions.

Case Report

Mrs. M. aged 25 years was admitted to L.L.R. Hospital (E.N.T. Department), Kanpur, on 6 March 1967 complaining of left-sided headache and excessive lacrimation (left eye) of one year duration and protuberance of left eye ball which was gradually becoming more and more prominent for the last 3 months. There was no history of nasal obstruction, nasal discharge, epistaxis or diplopia. She gave a history of an operation for similar complaints 6 years previously after which she remained symptom free for about 5 years. The previous operation notes were not available, but the reference card recorded fibrous dysplasia left ethmoid. Examination of the patient revealed left-sided proptosis with pushing of eyeball laterally and upwards and there was a scar medial to the inner canthus of left eye. Oedema of the eyelids (left) was present and on pressing over the lacrimal sac, thick discharge came out through lacrimal punctum. There was no swelling visible or palpable over the maxillary sinus or medial to the inner canthus of eye. Anterior rhinoscopy showed the middle meatus to be unobstructed. Posterior rhinoscopy showed the posterior end of middle turbinate oedematous. The lateral wall of nasopharynx (left side) was pushed medially. Ears, throat and larynx were normal. The patient’s hearing was normal and central nervous system examination, including fundus examination and field of vision, did not reveal any abnormality. Routine blood and urine examination were normal. X-ray of the paranasal sinuses revealed haziness of both maxillary antra, more marked on the left side and expansion of the medial wall of the left orbit with alternating areas of density and radiolucency (Figs. 1 and 2).

On 8 March 1967, the patient was operated under general anaesthesia. Howarth incision was used. Periosteum was elevated and anterior ethmoid cells removed; upper part of frontonasal process of maxilla and part of nasal bone were removed to get wide exposure. Firm, friable, bony mass was felt in the region of the medial wall of orbit and it was removed completely. Optic nerve was exposed while so doing. Frontal sinus opened and found healthy. Sphenoid
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sinus was opened and found to be full of hypertrophic mucosa which was removed. Posterior end of middle turbinate was also removed. Left maxillary antrum was opened by sub-labial approach and was found to be healthy. Incision wound closed.

Histopathological examination of the specimen removed showed normal looking bony trabeculae with loose oedematous, well-vascularized connective tissue in between and areas of necrosis and degeneration at places and hyalinization in other places, the picture suggesting fibrous dysplasia (Figs. 3 and 4).

The post-operative period was uneventful and patient was discharged on 20 March 1967. The proptosis completely disappeared even though patient still had epiphora. Anterior rhinoscopy showed the operative cavity to be completely healed (Fig. 5). Six months' follow-up did not show any recurrence of disease or symptoms.

Discussion

Fibrous dysplasia of bone is a polyostotic or monostotic condition, apparently due to a perverted activity of the specific bone-forming mesenchyme. It is generally agreed by most authors (Lichtenstein, 1938; Mallory, 1942; Schlumberger, 1946 and Szanto, 1952), based on clinico-pathological grounds, that ossifying fibroma is a variant of monostotic fibrous dysplasia. According to Szanto (1952) the only difference that exists is that in ossifying fibroma, the spherical islets of osteoid tissue predominate over the bony spicules, so characteristic of monostotic fibrous dysplasia. Smith and Zavaleta (1952) consider monostotic fibrous dysplasia of cranial or facial bones as synonymous with ossifying fibroma. Other authors, however, are strongly opposed to the opinion of identity of these conditions and consider monostotic fibrous dysplasia as a separate clinical entity (Thomas, 1956; Weinmann and Sicher, 1955).
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FIG. 5. Patient six months after operation.

Aetiology
The cause of this condition is not known. One view is that it is a true neoplasm. The second theory is that the lesion represents an abnormal reaction to trauma of the bone. It is maintained that after an injury, small haemorrhages occur within the bone, followed by an abnormal process tending to restoration of the lesion. This process causes a dissolution of the osseous tissue by osteoclastic cells and its replacement by fibrous connective tissue which has a slow and abnormal tendency to ossification. The local predisposition of the tissues is a factor to this process.

It has been suggested that the disease appears to be due to a developmental defect in the normal maintenance of the postnatal intramedullary bone. In the normal process of bone growth and maturation, mature bone undergoes physiological lysis and is replaced by solid masses of proliferating fibrous tissue cells. This process results in expansion and weakening of the cortex with resultant deformity.

Pathology
Macroscopically, the lesion may be white in colour and varies from softness to firmness in consistency. Some areas are gritty because of bony spicules in the fibrous tissue. Microscopically, spicules of osteoid tissue or calcified bone trabeculae are embedded in a moderately or markedly vascular fibroblastic...
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tissue; there are marked variations of the cellularity of fibrous tissue from area to area with occasional hyaline cartilagenous tissue, haemorrhages and cyst formation.

Clinical Features

The disease usually manifests itself in late childhood and adolescence and is supposed to be commoner in females. These lesions are painless, slowly growing non-invasive tumours that displace the surrounding structures. When occurring in the maxilla or ethmoid, they can produce nasal obstruction, proptosis and obvious swelling of face. The lesion probably begins before the growth period ends, and progresses rapidly during its first 5 years, more slowly afterwards. There is a tendency to gradual deceleration of growth so that the condition may become stationary in adult life, but slowing of the growth may not coincide with the termination of growth in the body skeleton.

Diagnosis and Management

Radiological study is of great value in the diagnosis. Fries (1957) has divided the lesions of the skull and facial bones into three categories based on radiological findings.

(i) Pagetoid type: The involved area is expanded and has alternate areas of density and radiolucency. The skull may become quite thickened. Most of the expansion is outward with thinning, erosion and disappearance of the outer table.

(ii) Sclerotic form: This type will appear homogenously dense radiologically.

(iii) Cyst-like form: Mostly seen in skull bones; round or oval in shape and it has a dense border which is thin and distinct.

Our case fits into the first variety, the pagetoid type. X-ray examination reveals distinctly or vaguely the existence of a tumour, but this is not enough for differential diagnosis. For this reason, it is important to perform a biopsy.

The only treatment in these cases is surgical excision of the tumour and complete curettement of the affected bone. This treatment usually prevents recurrence. These are benign and not metastasizing lesions. Malignant transformation has not been reported in the available literature.

Summary

A case report of fibrous dysplasia of the ethmoid is presented. A critical review of literature has been made with discussion on the aetiology, pathology, clinical features, diagnosis and management of these cases.

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REFERENCES


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