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Case Report

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Fibrous Dysplasia of the Orbit in an Infant

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Fibrous dysplasia · Orbit · Proptosis

Abstract
Fibrous dysplasia is an idiopathic fibro-osseous lesion of the skeletal bones. These uncommon osseous lesions usually manifest within the first two decades of life. Its occurrence during infancy is extremely uncommon, and we describe here a case of fibrous dysplasia involving the orbit in a 9-month-old male child.

Introduction

The fibro-osseous lesions involving the craniofacial bones include fibrous dysplasia, ossifying fibroma, osteoma and osteoblastoma. Fibrous dysplasia is regarded as an idiopathic disorder of the skeletal bone in which medullary bone is replaced by fibro-osseous tissue. This is histologically characterized by the replacement of spongy osteoid by fibrous tissue often showing varying degrees of maturity with woven bone in between them. This entity constitutes about 2.5% of all bone tumours [1]. Two distinct clinical manifestations of fibrous dysplasia are well-recognized, viz. polyostotic and monostotic variants. Fibrous dysplasia has a predilection to involve membranous bones such as maxilla, mandible, frontal and parietal bones [2]. Among the craniofacial bones, orbital bone forms the commonest site. The commonest clinical presentation of fibrous dysplasia involving orbital bone is proptosis and impairment of vision due to compression of the intraorbital part of the optic nerve. In these patients, the clinical manifestations usually occur in the first two decades. The review of Anglo-American literature did not reveal any documented cases during infancy. The purpose of this case report is to describe the occurrence of solitary fibrous dysplasia in a 9-month-old male child.

Case Report

A 9-month-old male child presented with unilateral painless, gradually progressing proptosis involving the right eye for the past one month. There was no loss of vision. The child was born of a full-term normal delivery of non-consanguinous parentage. The general physical examination was essentially normal except for the presence of umbilical hernia and pectus carinatum deformity of the chest. Neither any dermatological nor any endocrine disorders were present. Neurological examination showed an alert, well-oriented,

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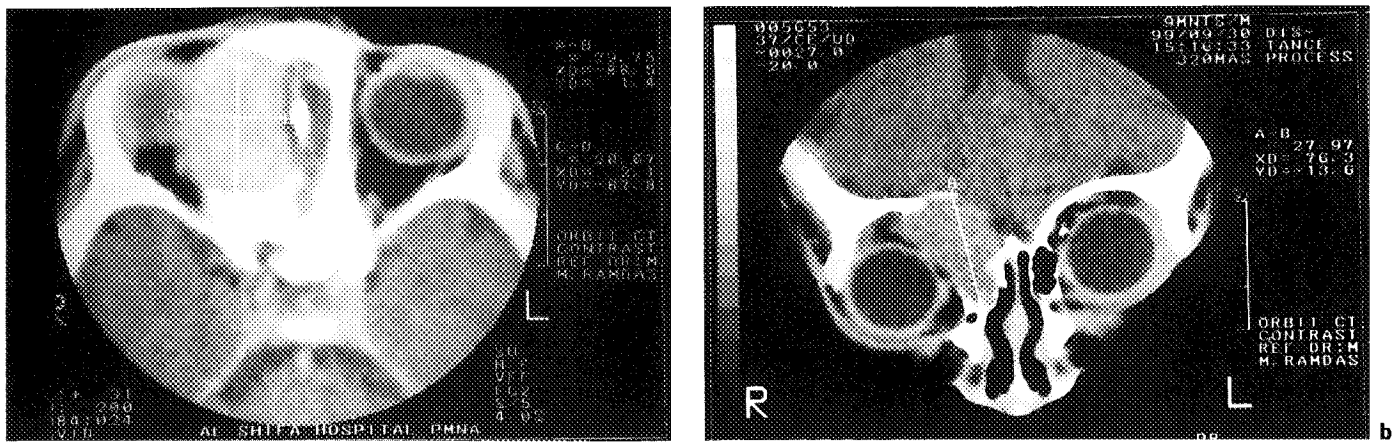

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Fig. 1. CT scan with contrast administration; axial (**a**) and coronal view (**b**) showing well-defined mildly enhancing extraconal lesion in the superomedial compartment of the orbit with erosion of the superomedial wall.

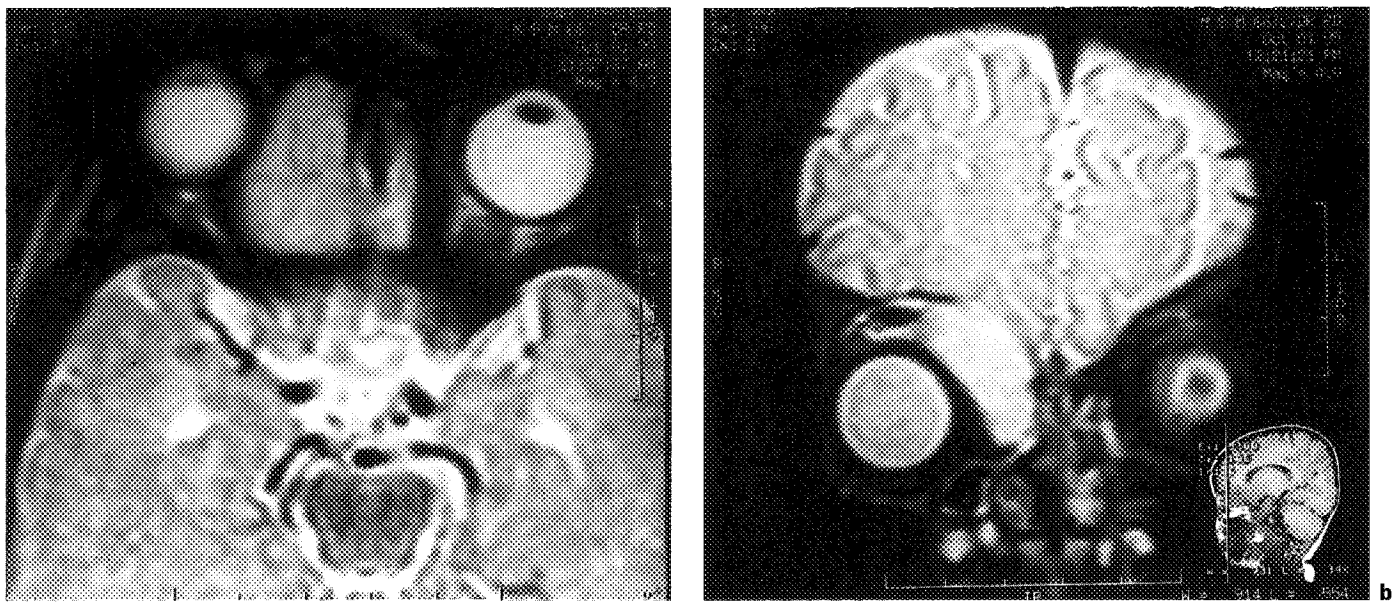

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Fig. 2. MRI scan, T_2-weighted images; axial (**a**) and coronal view (**b**) showing mildly hyperintense lesion in the same location.

co-operative child, and there were no focal neurological deficits. An ophthalmologic examination revealed an extra-axial painless mass involving the right eye. There were neither chemosis nor increased vasculature of conjunctiva. The eyeball was pushed laterally and inferiorly. The extraocular movements were well preserved. There was no nystagmus. The visual acuity and visual fields were normal in the right eye.

The routine laboratory investigations were within normal limits. The ultrasound scanning did not reveal any visceral abnormality. The computed tomography (CT) scan of the cranium showed a hyperdense mildly enhancing extraconal lesion in the superomedial compartment of the right orbit (fig. 1a, b). The medial rectus muscle was compressed and displaced laterally. The right optic nerve was normal in configuration. The lesion was seen medially extending into

the right ethmoid sinus and to the anterior cranial fossa. The lesion also was causing destruction of the superomedial wall and roof of the orbit. The magnetic resonance imaging (MRI) revealed the same lesion, which was isointense on T_1-, and hyperintense on T_2-weighted images (fig. 2a, b). The pre-operative impression was a metastatic lesion, which could be due to either neuroblastoma or leukaemic deposits.

The patient was subjected to a right supraorbital craniotomy. Peroperatively, there was a large greyish white firm vascular tumour in the superomedial compartment of the orbit causing bony destruction of the roof and medial wall of the orbit. The lesion was not attached to the periorbita or dura. The destroyed bone was removed and subtotal excision of the lesion was done. The lesion did not involve suture lines at the skull base. Orbital reconstruction was not required because of very small defect seen in the supraorbital margin. The surgical specimen was sent for routine histopathological examination. The postoperative period was uneventful, and the child was discharged on the eighth postoperative day. At 2 months follow-up, apart from a mild degree of proptosis of the right eye, ophthalmological and neurological examination were within normal limits.

Grossly, the specimen consisted of multiple irregular grey-white soft tissues along with several pieces of bone. The specimen was processed for histopathological studies in the routine protocol and examined with haematoxylin and eosin stain. Histologically, the lesion was composed of spicules of woven bone separated by fibrous tissue (fig. 3). The fibrous tissue showed varying degrees of cellularity. Cellular anaplasia and mitotic figures were absent. Neither inflammatory infiltrate nor haemorrhages within the lesions were seen. These histopathological features were interpreted as fibrous dysplasia. Histopathologically no other differential diagnosis was considered.

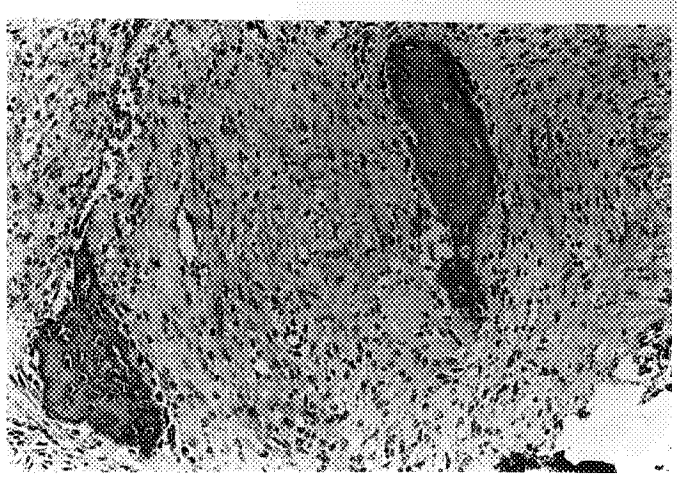

Fig. 3. Photomicrograph showing woven bone seperated by fibrous tissue having a moderate degree of cellularity. HE. × 180.

Discussion

Fibrous dysplasia is a benign fibro-osseous lesion of unknown actiology involving skeletal bones. Fibrous dysplasia can occur as monostotic or polyostotic forms, the latter may be associated with multiple endocrinopathies. 20% of monostotic and 40–60% of polyostotic cases may involve craniofacial bone [3]. The characteristic 'lion face' or 'Leontiasis Ossea' may develop if the facial bones are extremely involved in the disease processes. Bone expansion may encroach the paranasal sinuses and orbit [4]. Facial abnormalities were not observed in our case.

Henderson [5] in a series of 764 orbital tumours at the Mayo Clinic over a 16-year period, recorded only 5 patients with fibrous dysplasia. Jeffrey et al. [6] reported a 3-year-old boy with extreme fibrous dysplasia involving the body of the sphenoid bone, who presented with loss of vision. The patients with craniofacial fibrous dysplasia can present with acute visual loss [7]. Bilateral proptosis as a presenting feature of orbital fibrous dysplasia has been reported by Horgan et al. [8]. In our case, there was no visual impairment, and the patient presented with only unilateral proptosis.

Clinically, the fibrous dysplasia is often reported to occur in the first two decades of life, and the bony lesion may remain static in growth after puberty [2]. The recurrence in adulthood form of fibrous dysplasia has been reported in about 37% of cases [9]. Jan et al. [10] reported a case of fronto-orbital fibrous dysplasia in an 8-year-old boy. The review of the literature failed to reveal any case of fibrous dysplasia in the first year of life. To the best of our knowledge, the case reported here is the youngest age for fibrous dysplasia occurring in orbit.

The radiological appearances of fibrous dysplasia of the craniofacial bone may range from very radiolucent lesions composed of mainly fibrous tissue to markedly sclerotic expanded bone made up of dense bony trabeculae with little fibrous element. Any degree between these two extremes may be present and thus fibrous dysplasia mimics a wide range of other pathologies of craniofacial bones. The neuroradiological differential diagnosis includes psammomatoid ossifying fibroma, osteoblastoma and meningioma.

Aside from the sclerotic and cystic forms of fibrous dysplasia already described in the literature, Jan et al. [10] reported a flesh tumoral form involving the left optic canal. The lesion belongs to the compact type in our case. Microscopic appearance of this entity is unique and is composed of cellular fibrous connective tissue separated by woven bone. This histological feature may mimic ossifying fibroma, but is composed of lamellar bone.

The surgical management of fibrous dysplasia is curative. Although the histological appearance is benign, rare-

ly malignant transformation to osteosarcoma, chondrosarcoma and malignant fibrous histiocytoma is reported [11, 12]. Accurate histopathological interpretation is essential because these lesions are benign and do not require any post-operative radiotherapy.

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