Case Report

Fibrous Dysplasia of the Orbit in a 5-Year-Old Girl

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Abstract

Purpose: The aim of the present study was to describe a case of orbital fibrous dysplasia in a very young child.

Case report: A 5-year-old girl presented with unilateral, painless, slowly progressive proptosis of the left eye and inferotemporal globe displacement from about 1 year ago. The visual acuity in both eyes was 20/20. The general physical examination was normal. Computed tomography (CT scan) revealed thickening and deformity of the orbital walls. Lateral orbitotomy and biopsy were performed and were compatible with fibrous dysplasia.

Conclusion: Craniofacial fibrous dysplasia usually occurs in the second decade of life, but it can also rarely present in the early years of life. Therefore, fibrous dysplasia should be considered as a differential diagnosis of proptosis or globe displacement in the early years of life.

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Introduction

Fibrous dysplasia (FD) is a benign slowly progressive disorder of bone, where normal cancellous bone is replaced by fibrous tissue and immature woven bone. This entity constitutes about 2.5 % of all bone tumors [1].Two clinical forms of FD are recognized: polyostotic and monostotic variants. Polyostotic lesions combined with cutaneous pigmentation, precocious puberty and endocrine dysfunction is called Albright's syndrome [2]. FD has a predilection to involve membranous bones such as maxilla, mandible, frontal and parietal bones. When FD occurs in the craniofacial region, orbits are commonly involved [3]. Patients with orbital FD usually present with classical manifestations of proptosis, diplopia and periorbital facial asymmetry [4]. Other ocular complications have been reported including exposure keratitis, displacement of the globe and refractive errors secondary to compression and distortion of globe. If the sphenoid bone is involved, the reduced diameter of the optic canal can lead to optic nerve damage and visual loss [5]. In this manuscript we report a 5-year-old child with orbital fibrous dysplasia.

Case report

The present report was approved by the ethics committee of Baqiyatallah University of Medical Sciences and informed consent was given by patient's parents for reporting this case. A 5-year-old girl presented with unilateral, painless, gradually progressive proptosis of the left eye from about 1 year ago. The visual acuity of both eyes was 20/20.The globe was displaced inferotemporally, but there was no ocular motility restriction or diplopia (Figure 1).The general physical examination was normal.

Computed tomography (CT-Scan) of the orbit revealed deformity of the orbital walls and thickening of the large wing of the sphenoid, squamous part of the temporal bone, zygomatic arc and frontal bone. The left optic canal and optic nerve were normal in configuration. Paranasal sinuses had normal configuration and pneumatization (Figure 2).

Lateral orbitotomy and biopsy were performed. The surgical specimen was sent for routine histopathological examination. The postoperative period was uneventful.

Grossly, the specimens consisted of multiple irregular creamy-gray tissue with bony consistency. Histologically, the lesion was composed of fragments of trabecular bone with reactive change, beside small pieces of neoplastic tissue composed of woven and trabeculae bone in fibrous stroma. These histopathological findings were compatible with fibrous dysplasia.

Discussion

Von Recklinghausen described a number of cases whose normal bones were replaced by immature bone and fibrous tissue in 1891, and the term fibrous dysplasia was first used in 1938 by Lichtenstein [5]. FD is a benign fibro-osseous lesion of unknown etiology involving skeletal bone and can occur as monostotic or polyostotic forms.

Monostotic FD is about 4 times more frequent than its polyostotic form [6]. Craniofacial bones

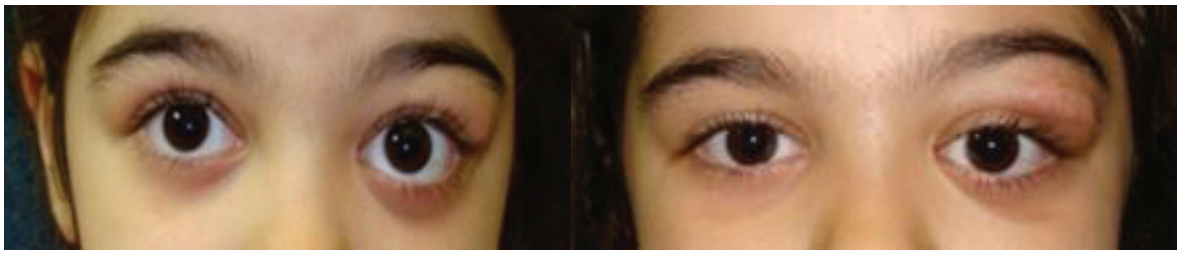

Figure1: Left eye proptosis and inferotemporal displacement of the globe, before biopsy (Left) and after biopsy (Right).

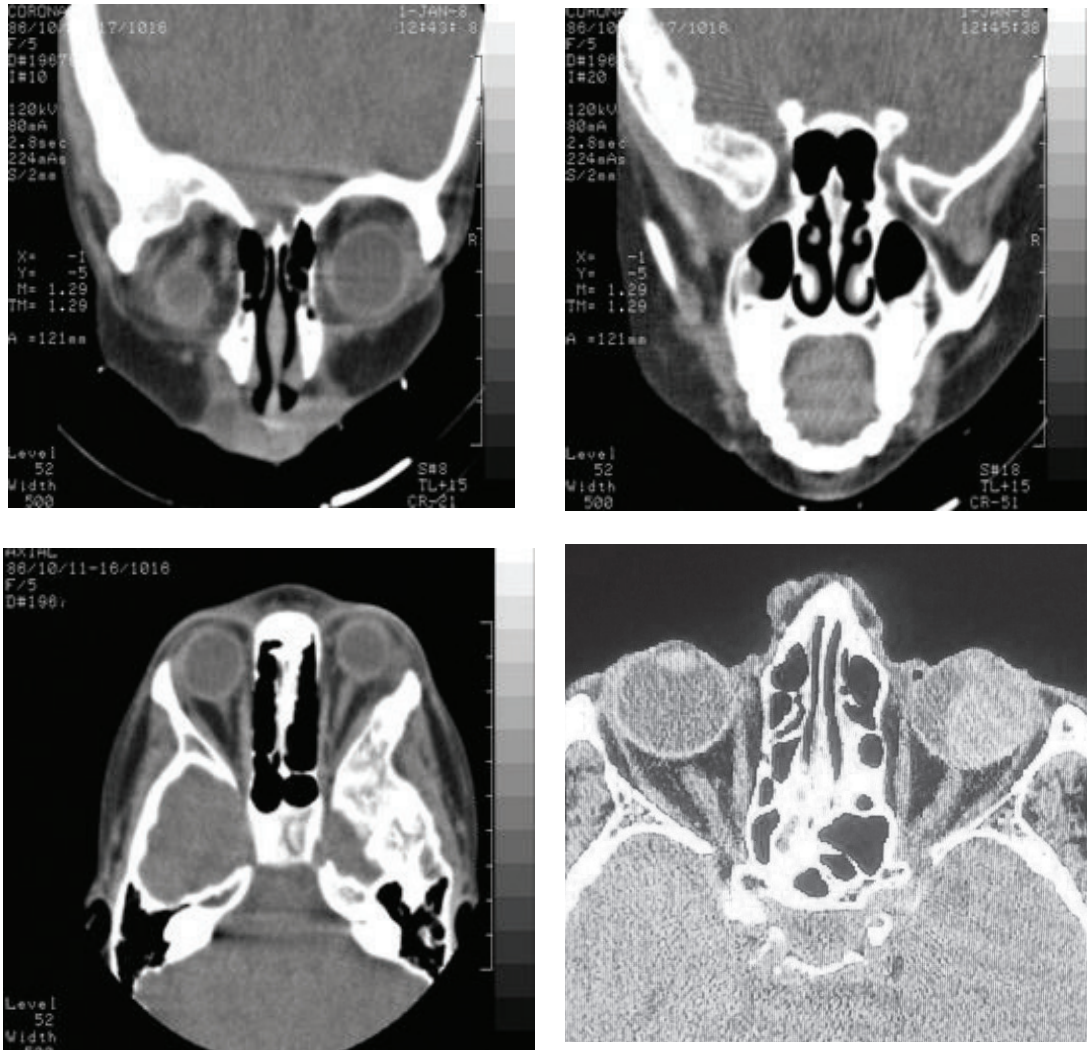

Figure 2: Deformity and thickening of the orbital walls and inferotemporal displacement of the globe (Coronal view) and mild proptosis (Axial view).

may be involved in 20 % of monostotic and 40 - 60 % of polyostotic cases [7].

FD generally occurs during the second decade of life and lesions may remain static in growth after puberty [8,9]. In this study we reported an unusual case of FD in a 5 year-old-girl. Fadle et al., reported 22 cases with orbitocranial FD and the youngest patient in their study was 17-year-old (mean age was 29.5) [10]. Leong et al., reported a 12 year-old-girl with craniofacial FD with orbital involvement, who developed progressive visual impairment [11].

Visual manifestations are common in craniofacial FD and include proptosis, which is present in about 30 % of cases, globe displacement and strabismus, diplopia, extraocular muscle palsies, epiphora and visual loss due to optic nerve compression [5,6]. The presenting symptoms in our case were mild proptosis and globe displacement that began in the 4th year of life.

The radiological appearance of FD may range from very radiolucent lesions composed of mainly fibrous tissue to markedly sclerotic expanded bone made up of dense bony trabeculae. Any degree of appearance between these two extremes may be present [3].

Treatment of FD includes surgical and non-surgical options, but the results of nonsurgical

options are poor. Indications for surgery are both cosmetic and functional [6]. Complete resection of the involved area is the treatment of choice, but this invasive approach may contribute to functional impairments and secondary deformities that are more damaging for the patient than the FD itself [12]. There is little risk of malignant degeneration, with transformation of dysplastic bone to osteogenic sarcoma, fibrosarcoma and chondrosarcoma [5]. The incidence of malignant transformation in normal bone is 0.001 % but in FD it is 400 times greater [5]. Radiotherapy may increase the risk of malignant transformation [2].

Conclusion

Craniofacial fibrous dysplasia usually occurs in the second decade of life, but it can also rarely present in the early years of life. Therefore, fibrous dysplasia should be considered as a differential diagnosis of proptosis or globe displacement in the early years of life.

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Footnotes and Financial Disclosures

Conflict of Interest:

The authors declare no conflict of interest with the subject matter of the present manuscript.