(49)

JOURNAL OF MEDICAL CASE REPORTS

Case report

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Visual impairment from fibrous dysplasia in a middle-aged African man: a case report

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Journal of Medical Case Reports 2009, **3:**14 doi:10.1186/1752-1947-3-14

The electronic version of this article is the complete one and can be found online at:
http://www.jmedicalcasereports.com/content/3/1/14

Received: 28 May 2008
Accepted: 13 January 2009
Published: 13 January 2009

Abstract

Introduction

Fibrous dysplasia is a benign tumour of the bones and is a disease of unknown aetiology. This report discusses a case of proptosis and visual deterioration with associated bony mass involving the right orbit.

Case presentation

A 32-year-old Nigerian man of Yoruba ethnic origin presented to the eye clinic of our hospital with right-eye proptosis and visual deterioration of 7-year duration. Presentation was preceded by a history of trauma. Proptosis was preceded by trauma but was non-pulsatile with no thrill o associated with bony orbital mass. The patient reported no weight loss. Examination of his right eye show 6/60 with relative afferent pupillary defect. Fundal examination revealed optic atrophy. Computed tomogr expansile bony mass involving all the walls of the orbit. The bony orbital mass was diagnosed histological dysplasia. Treatment included orbital exploration and orbital shaping to create room for the globe and rel the optic nerve.

Conclusion

Fibrous dysplasia should be considered in the differential diagnosis of slowly developing proptosis with as: in young adults.

Introduction

Fibrous dysplasia is a benign, slowly growing disorder of bone in which the normal cancellous bone is repl woven bone and fibrous tissue [1]. This condition was first reliably recognized by von Recklinghausen in 1 then, a large number of cases have been reported and considerable advances have been made in the unc treatment of the disease [3] which constitutes 2.5% of all bone tumour and 7.5% of all benign bone neop no sex preference [3] and usually manifests before the 3rd decade of life [5]. Fibrous dysplasia has two b namely the monostotic and the polyostotic forms [3]. The monostotic form of this disease constitutes abc and only involves the craniofacial skeleton in about 10% of cases, having a predilection for the ribs and fe

Histological examination provides the basis for an accurate diagnosis. The tumour is characterized by mul irregular spicules of immature bone superimposed on a background of moderately cellular fibrous connect However, ancillary investigations, like computerized tomography (CT) which shows the characteristic 'gro appearance in the sclerotic form and non-homogenous appearance in the cystic and mixed form, may be complement findings of histopathology.

Fibrous dysplasia may cause ophthalmic problems such as proptosis and dystopia, ocular motility problem deformity; however, visual loss represents the most common neurological complication of fibrous dysplas skull [7].

Fibrous dysplasia, though not rare, is a disease mainly documented among Caucasians [1,3,8] and Asians are found in the literature of this problem among African Nigerians [10]; especially of the monostoic form orbital involvement.

Case presentation

A 32-year-old Nigerian businessman of Yoruba ethnic origin was referred to the eye clinic of our hospital f hospital in Nigeria with complaints of progressive protrusion of the right globe for 7 years. Six months pri his complaints, he had hurt the edge of the right superior orbital margin against the edge of an iron bed a No treatment was received for this. The protrusion of the globe continued to increase for about 7 months There was no pain and no diplopia but there had been progressive deterioration of the vision in the eye. I of the right orbit performed at the referring hospital revealed a bony hard mass involving both the lateral wall. This mass could not be removed. There was no history of weight loss, heat intolerance or excessive proptosis was not made worse by the Valsalva manoeuvre. The patient experienced no unusual noises in no history of swelling (of bony or soft tissue) in other parts of the body. Furthermore, there were no hoar dysphagia, cough, palpitation, headache, vomiting or seizure and no focal neurological deficits. The patier hypersensitivities and no diabetes or asthma. He was single, the first of 6 children of his parents. His fath age of about 62 years of an unknown cause, the mother was alive and well, aged about 60 years. There v history of similar eye problems.

Examination revealed an otherwise healthy-looking man, with normal systemic examination. The right oc revealed a visual acuity of 6/60 with a proptosis of 17 mm (Hertel exophthalmometer). The proptosis was (inferotemporally), non-retropulsive, nonpulsatile, nontender and had no thrill and no bruit. There was ch overlying conjunctiva with moderate restriction of the extra-ocular muscle movement in all direction of th pupillary reaction was sluggish with a relative efferent pupillary defect. Fundoscopy showed a pale disc wi margins. The left eye had a visual acuity of 6/5 with normal anterior and posterior segments. Cranial com scan (Figures 1 and 2) showed a right expansile bony mass involving 1) the orbital roof and especially the the sphenoid; and 2) the medial and lateral walls of the orbit, especially the greater wing of the sphenoid inferior encroachment involving the right retro-orbital space with compression of the globe against the m No intracranial extension was seen. A clinical diagnosis of fibrous dysplasia was made.

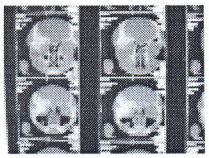

Figure 1. Preoperative cranial computed tomography scan showing expansile bor mass of the right orbit involving the greater wing of the sphenoid, the medial wal the orbit and the greater wing of the sphenoid (frontal view). There is encroachmer into the retro-orbital space with obvious proptosis of the right globe. There is no evidence intracranial extension.

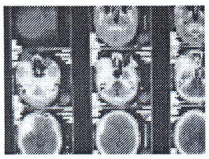

Figure 2. Preoperative cranial computed tomography scan showing expansile bor mass of the right orbit involving the greater wing of the sphenoid, the medial wal the orbit and the greater wing of the sphenoid (transverse view). There is encroachment into the retro-orbital space with obvious proptosis of the right globe. There evidence of intracranial extension.

The ophthalmic, neurosurgical and maxillofacial units of our hospital carried out a joint surgical exploratic through a modified lateral orbitotomy, using an electric drill for the lateral orbital wall. Operative findings thickened expanded zygomatic bone and greater wing of sphenoid, with the orbit being almost completel expanded dense bony tissue. Gradual removal of the bony mass was performed in layers, using a hamme new orbital space was created. The histological finding (Figure 3) was in keeping with fibrous dysplasia ar broad sheets of interconnection trabeculae of calcified bone with sparsely cellular intervening vascular co stroma. Postoperatively, there was reduction in the degree of proptosis. The patient was discharged after days at which time the proptosis had reduced to 6 mm (Hertel exophthalmometer). At the time of writing visual acuity remains at 6/60 and he is receiving follow-up in the outpatient clinic.

Figure 3. Histology photograph of specimen obtained at surgery, showing broad sheets of trabeculae of calcified bone with sparsely cellular intervening connectiv tissue stroma.

Discussion

Fibrous dysplasia results from a defect in osteoblastic differentiation affecting the final maturation of the Although described as a non-familiar, congenital disorder of the bone, it usually manifests before the 3rd Our case fell within the age group described in the literature. The history of trauma preceding the onset c this case may be of interest. This is because there had also been a few reports describing a cause-and-ef between fibrous dysplasia and trauma [12]. However, the 'bumping into objects' described by the patient -existing visual impairment or field defect in the affected eye which was not noticed by the patient until t trauma; even more so as there was no objective visual acuity or field assessment prior to the period. Tra puberty when bone development is at its maximum may have implications on the development of tumour this may be difficult to establish in this case as bony growth should have concluded prior to the age when trauma.

The rapid worsening of visual acuity as described in this case could be as a result of cyst formation within resultant compression of the optic nerve and impairment of the venous return from the orbit. This is supp that there was lot of conjuctival chemosis which resolved after surgical decompression of the orbit. The lc may also have been a result of haemorrhage into the tumour resulting from the trauma sustained. Howe be substantiated from the histology results. Visual impairment following fibrous dysplasia has been attrib factors which include optic-nerve traction due to proptosis, sinus mucocele formation with raised intra-orl haemorrhage within the tumour, optic canal stenosis, as well as cyst formation within the lesion [13].

Establishing the diagnosis of fibrous dysplasia requires close cooperation between clinician, radiologist an which was demonstrated very well in the case reported. Orbital osteoma which is the most common beni paranasal sinuses [14] may at time present a diagnostic challenge. This is occasioned at times by the nor histological and radiological appearance which may result in poor characterization of the lesion.

Therapeutic indication depends on the course of tumour and the development of complications. This coul observation with serial radiological follow-up to medical therapy with systemic corticosteroid and surgical surgical option adopted in this case met with the basic tenets of operative intervention using the treatme proposed by Chen and Noordhoff [15]. There was an obvious neurological effect as demonstrated by the reduction in the visual acuity as well as the cosmetically unacceptable degree of proptosis; even more so young man. Although there was no visual improvement postoperatively despite postoperative administrat corticosteroid, this may not be surprising because of the large interval between the onset of symptoms a surgical orbital decompression. However, part of the patient expectation was met as shown by the reduct of proptosis.

Complete resection of the lesion was not possible in this case because the entire posterior orbit was filled We concentrated on a curettage to provide enough room for repositioning of the globe, using a lateral ort which is associated with less morbidity and quick recovery. Cranio-orbital shaping is an acceptable mode treatment for fibrous dysplasia when it may not be possible to remove the pathological bone completely.

Conclusion

Monostotoic fibrous dysplasia of the orbit causing neuro-ophthalmic complications associated with compre should be considered in the differential diagnosis of slowly progressive proptosis in young adults.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

COB took part in the surgery and was a major contributor to preparing the manuscript. AOF, JKC and VN surgery. OOK took part in the surgery and was a major contributor to preparing the manuscript. AOO per histological examination of the specimen. AIA and AMB were major contributors to the manuscript prepar read and approved the final manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompan copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

1. Bibby K, McFodzean R: **Fibrous dysplasia of the orbit.**
Br J Ophthalmol 1994 , **78:**266-270. PubMed Abstract | Publisher Full Text | PubMed Central Ful
Return to text

2. von Recklinghausen FD: **Die Fibrose oder deformierende Ostitis, die Osteomalacie und di Carcinose in ihren gegenseitigen Beziehungen.** In *Festschrift Rudolf Virchow zum 13. Oktol* Georg Reimer Verlag; 1891.
Return to text

3. Ricalde P, Horswell BB: **Craniofacial fibrous dysplasia of the fronto-orbital region: a case literature review.**
J Oral Maxillofac Surg 2001 , **59:**157-168. PubMed Abstract | Publisher Full Text

Return to text

4. Edgerton MT, Persing JA, Jane JA: **The surgical treatment of fibrous dysplasia. With emph contributions from craniomaxillo-facial surgery.**
Ann Surg 1985 , **202:**459-479. PubMed Abstract | Publisher Full Text | PubMed Central Full Text
Return to text

5. Finney HL, Roberts TS: **Fibrous dysplasia of the skull with progressive cranial nerve invo**
Surg Neurol 1976 , **6:**341-343. PubMed Abstract
Return to text

6. Hoffman S, Jacoway JR, Krolls SO: **Fibrous dysplasia: Benign nonodontogenic tumors of th**
Intraosseous and Periosteal Tumors of the Jaws. 2nd edition. Edited by: Seymour Hoffman MD. Forces Institute of Pathology; 1987:211-216.
Return to text

7. Sassin JF, Rosenberg RN: **Neurological complications of fibrous dysplasia of the skull.**
Arch Neurol 1968 , **18:**363-369. PubMed Abstract
Return to text

8. McCluskey P, Wingate R, Benger R, McCarthy S: **Monostotic fibrous dysplasia of the orbit: a lacrimal fossa mass.**
Br J Ophthalmol 1993 , **77:**54-56. PubMed Abstract | Publisher Full Text | PubMed Central Full T
Return to text

9. Panda A, Dayal Y, Vasistha S, Patnaik NK: **Fibrous dysplasia of the orbit.**
Indian J Ophthalmol 1985 , **5:**317-319.
Return to text

10. Odeku EL, Martinson FD, Akinosi JO: **Craniofacial fibrous dysplasia in Nigerian Africans.**
Int Surg 1967 , **51**(2)**:**170-182. PubMed Abstract
Return to text

11. Riminucci M, Fisher LW, Shenker A, Spiegel AM, Bianco P, Gehron Robey P: **Fibrous dysplasia McCune-Albright syndrome: abnormalities in bone formation.**
Am J Pathol 1997 , **151:**1587-1600. PubMed Abstract | PubMed Central Full Text
Return to text

12. Schlumberger HC: **Fibrous dysplasia of single bones (monostotic fibrous dysplasia).**
Mil Surg 1947 , **99:**504-527.
Return to text

13. Liakos GM, Walker CB, Carruth JA: **Ocular complications in craniofacial fibrous dysplasia.**
Br J Ophthalmol 1979 , **63:**611-616. PubMed Abstract | Publisher Full Text | PubMed Central Ful
Return to text

14. Selva D, White VA, O'Connell JX, Rootman J: **Primary bone tumors of the orbit.**
Surv Ophthalmol 2004 , **49:**328-342. PubMed Abstract | Publisher Full Text
Return to text

15. Chen YR, Noordhoff MS: **Treatment of craniomaxillofacial fibrous dysplasia: how early an extensive?**
Plast Reconstr Surg 1991 , **87:**799-800. PubMed Abstract
Return to text

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