FIBROUS DYSPLASIA OF THE ORBIT

A CASE REPORT

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Conflict of interest: None

INTRODUCTION

- Fibrous dysplasia is a non-neoplastic bone disorder associated with a defect in differentiation and maturation of osteoblasts
- As a result of this defective process, normal bone is replaced with fibrous tissue which leads to bone weakness and increased susceptibility to fractures
- Fibrous dysplasia is most commonly diagnosed in children and young adults before the age of 30 years
- It is usually sporadic, however association with endocrinopathies, such as McCune-Albright syndrome, has been reported
- Fibrous dysplasia is associated with GNAS gene mutation which affects osteoblast precursors as well as melanocytes and endocrine cells

AIMS: To report a case of orbital fibrous dysplasia in an adult patient who presented with mild unilateral relative proptosis

METHODS: Female patient with mild unilateral relative proptosis underwent full neuro-ophthalmic assessment with a CT and MRI of her head and orbits

RESULTS I

- A 55-year-old female presented to the eye clinic with 6-month history of mild swelling around her right eye and right eye looking more prominent compared to the left
- Patient's past medical history included multiple sclerosis, bipolar affective disorder, complete thyroidectomy for invasive follicular carcinoma and subsequent radioiodine treatment
- On examination patient had VA 6/6 in both eyes, full colour vision bilaterally, mild right upper eyelid oedema, constant relative proptosis of 3mm on the right side, no lid lag or lid retraction, good retropulsion, full eye movements, pupils equal and reactive to light with no RAPD, normal IOP in both eyes
- Anterior and posterior segment examination was unremarkable
- Urgent MRI head and orbits was arranged to rule out orbital metastasis from the invasive thyroid follicular carcinoma
- MRI revealed sclerosis and expansion of the right superotemporal bony orbit, extending into the clinoid process and associated with displacement of the lateral rectus muscle as well as prolapse of the right lacrimal gland - it was felt that these radiological features could represent fibrous dysplasia

CONCLUSIONS

- Craniofacial fibrous dysplasia should be suspected in patients presenting with cranial or facial asymmetry, nasal congestion, proptosis or visual disturbances
- In a patient with a new onset proptosis and history of malignancy, imaging of the orbits is crucial to rule out potential metastasis
- Although follicular thyroid carcinoma is associated with lytic rather than sclerotic bone metastasis, sclerosis can occur following radioiodine treatment
- As fibrous dysplasia is a benign process and very rarely undergoes malignant transformation (<1%), treatment is only reserved for cases where function is being threatened and involves surgical resection of the sclerotic bone
- In case of pain associated with fibrous dysplasia, medical management is based on bisphosphonates

RESULTS II

- The case was subsequently discussed with radiologist and clinical oncologist. Since sclerosis of lytic bone metastasis can occur following radioiodine treatment, it was felt that metastasis from follicular thyroid carcinoma should be excluded
- Patient underwent whole-body scintigraphy which confirmed changes typical of fibrous dysplasia affecting the right orbital roof and lateral wall
- In view of mild symptoms and no threat to vision, patient did not require any treatment . She has been followed up by ophthalmology on a yearly basis and so far orbital fibrous dysplasia has been stable, both clinically and radiologically

Fig 1. CT head and orbits. Homogenous sclerosis of the right superotemporal orbit, extending into the clinoid process.

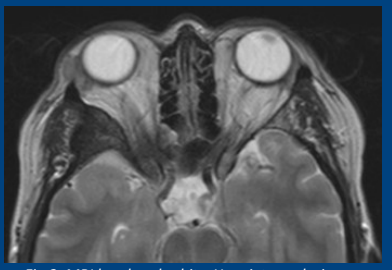

Fig 2. MRI head and orbits. Hypointense lesion of the right superotemporal orbit, causing displacement of the right lateral rectus muscle.

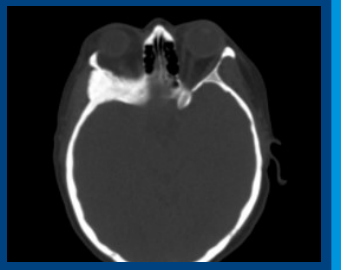

Fig 3. Bone scintigraphy. Increased uptake in the roof and lateral wall of the right orbit, which corresponds with an area of sclerosis.

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