Management of anterior skull-base fibrous dysplasia in children

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Introduction:

Fibrous dysplasia of the anterior skull-base can lead to severe facial disfigurement with optic nerve compression and blindness. The management of this condition in children can be controversial.

Objectives:

We evaluate presentation and outcomes in our case series of children with anterior skull-base fibrous dysplasia from 2011 to date in a single UK paediatric tertiary centre with a review of the available literature.

Background:

Fibrous dysplasia (FD) is a non-malignant condition in which normal bone and marrow are replaced by fibrous tissue and haphazardly distributed woven bone. ^[1] The craniofacial region is the most commonly involved site and the anterior cranial base is involved in >95% of cases of multi-bone (polystotic) disease. ^[2]

Asymmetry, facial pain and swelling are the most common presenting complaints. ^[3]

Rationale for bisphosphonate treatment:

Histologic findings often show high numbers of osteoclasts and evidence of bone resorption in FD lesions. Bisphosphonates improve the increased turnover of bone in FD by inhibiting osteoclast activity both directly and indirectly.

Radiological findings:

A classical 'ground-glass' appearance is described on CT imaging. ^[4]

Management:

This is challenging in the paediatric population due to continued bony skeletal growth, raised growth hormones during adolescence and potential side effects of medication used for symptom control in adults.

Authors recommendations:

1. Optimal care requires comprehensive evaluation and multi-disciplinary involvement with paediatric endocrinology/ ophthalmology and craniofacial teams.
2. Surgical treatment of stable disfiguring lesions should be postponed until skeletal maturity is complete [post-puberty wherever possible].
3. Surgical resection prior to skeletal maturity is recommended only if there is a significant risk to vision.
4. Potential use of adjuvant therapy such as bisphosphonates should be considered in all cases after discussion with paediatric endocrinology. Appropriate long term growth and biochemical bone markers surveillance necessary.

Results:

- 7 children in total.
- Age range at presentation: 3 -14 years [Mean = 7 years]
- 1 child had McCune Albright syndrome. The other children had monostotic disease and no other endocrine co-morbidity.

Symptoms:

- 5 children had complete unilateral visual loss
- 1 child had seizures
- 1 child had intractable facial pain

Management:

- 1 child underwent a clival biopsy. This was followed by rapid regrowth of diseased bone.
- 2 children required urgent transnasal endoscopic optic canal decompression for sudden visual deterioration with objective improvement in visual acuity.
- All 7 children were commenced on Bisphosphonate therapy under the guidance of paediatric endocrinology.

Follow-up:

- No further progression of the disease has been noted on subsequent imaging to date for any child.
- Surveillance bone density scans remain normal in all but the child with McCune Albright syndrome.
- Biochemical markers of bone turnover remain stable
- To date vision remains stable in the unilateral functioning eyes.

Discussion

A lack of evidence for Surgical management of fibrous dysplasia in children remains. The disease process is more active in childhood and the rate of regrowth post surgery is believed to be higher. There is a paucity of long term outcomes in children. Growth hormone excess has been shown to be associated with a higher rate of regrowth post surgery. Bisphosphonate therapy with surgery to decompress the remaining optic canal reserved for objective evidence of acute deterioration seems to have been an effective strategy in our patients.

References:

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