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A case of crouzon syndrome in a 46 year old female patient- A case report and study of literature

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Abstract

Case Report: A 46 yr old female presenting with defective vision of both eyes since childhood. A 46 yr old female, presented with defective vision both eyes since childhood, which has progressed gradually and attained the present appearance.

Past history: right side mastectomy done 10 yrs ago for carcinoma breast. She takes mixed diet, not a known diabetic and hypertensive. Family history nil relevant.
BP: 120/80 mm of hg PR: 78 / min.

Keywords: Crouzon syndrome, Proptosis, High arched palate

Introduction

Here we are presenting a case of crouzon syndrome in a 46 yr old female

CT Brain Done

Mid facial hypoplasia, mandibular prognathism, low set ears, high arched palate. On local examination: both eyes proptosed, left eye exotropia, bilateral lagophthalmos, cornea clear, conjunctival congestion, pupil ill sustained reaction, lenticular opacity.

Fundus both eyes: media: lenticular opacity present, optic disc pale with margins well defined, attenuation of vessels and sheathing, tessellation present.

Investigations: CT brain done.



Discussion

1- Crouzon syndrome is a craniosynostoses caused primarily by premature fusion of the coronal and sagittal sutures.

In which there is abnormally shaped skull

Inheritance is usually AD, The gene (FGFR2) has been isolated to chromosome 10.

Systemic features

- Short anteroposterior head distance and wide cranium due to premature fusion.
- Midfacial hypoplasia and curved 'parrot-beak' nose which gives rise to a 'frog-like' facies and mandibular prognathism.
- Inverted V-shaped palate.
- Acanthosis nigricans is seen in one subtype.

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Ocular features

- Proptosis due to shallow orbits is the most conspicuous feature.
- Hypertelorism (wide separation of the orbits).
- ‘V’ exotropia.
- Ametropia and amblyopia.
- Vision-threatening complications include exposure keratopathy and optic atrophy, due to chronic papilloedema and cerebral hypoperfusion secondary to sleep apnoea.

Ocular associations include blue sclera, cataract, ectopia lentis, glaucoma, coloboma, megalocornea and optic nerve hypoplasia.

- 2- Compression of nasal passages, often causing reduced airflow through the nose
- Large, protruding lower jaw
- Misalignment of teeth
- High-arched, narrow palate, or cleft palate

Other symptoms and complications that can result from crouzon syndrome include:

- Problems with development of the inner ear and hearing loss
- Meniere’s disease—lightheadedness, vertigo, or ringing in the ears
- Problems with the eyes, including vision problems, crossed eyes, or involuntary eye movement
- Curvature of the spine
- Headaches
- Acanthosis nigricans—small, dark, velvety patches of skin
- Hydrocephalus—buildup of fluid in the skull

- 3- In crouzon syndrome in addition to craniofacial synostoses there is syndactyly
- 4- Coarctation of aorta

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