Case Report

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Crouzon's Syndrome in 7 years old child: A Comprehensive Review and Clinico-Radiological Illustration of a Case.

Abstract:

Crouzon's syndrome is a rare genetic disorder, described by a French neurosurgeon in 1912. It is caused by a mutation in the fibroblast growth factor receptor 2 (FGFR2) gene. The disease is characterized by premature synostosis of coronal and sagittal sutures which begins in the first year of life. It occurs in approximately 1 in 25,000 births worldwide. Here, we report a case of this rare entity. The patient presented with brachycephaly, maxillary hypoplasia, exophthalmos, and mandibular prognathism along with dental and orbital abnormalities.

Key-words: Craniofacial dystosis, Crouzons syndrome, Copper beaten appearance, Exophthalmos

Introduction:

Crouzon syndrome (CS) is a rare genetic disorder characterized by premature closure of one or more cranial sutures and produces the characteristic craniofacial and other associated abnormalities.[1] Once the sutures close, the growth potential of those sutures is restricted. However, multiple sutural synostoses frequently extend to premature fusion of the skull base causing midface hypoplasia, shallow orbit, maxillary hypoplasia, and occasional upper airway obstruction.[2] Other clinical features include hypertelorism, exophthalmos, strabismus, beaked nose, short upper lip, maxillary hypoplasia, and relative mandibular prognathism.[3]

Crouzon's syndrome occurs in approximately 1 in 25,000 births worldwide. It makes up approximately 4.8% of all cases of craniosynostosis.[2] The differential diagnosis of Crouzon's syndrome includes simple craniosynostosis as well as Apert syndrome, Carpenters syndrome, Saethre-Chotzen syndrome, and Pfeiffer syndrome.[4] Crouzon syndrome is distinguishable from other craniosynostosis syndromes by lack of hand and/or foot abnormalities.[1] In this article, we present a case of Crouzon's syndrome in a 7-year-old child patient.

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Quick Response Code

Case report:

A 7-year-old boy reported to our department for treatment along with his mother. The chief complaint as presented by the mother was a pain in the child's upper right back region of jaw since 4-5 days. Pain was throbbing in nature and child was not able to chew on that side and aggravated after taking hot and cold. Since the child's appearance and head size were not normal, family and medical history were taken in detail. A review of medical history was unremarkable. The mother reported normal labour and delivery. The patient's mother had given the history of the patient's grandfather and after that patient's father and paternal aunt had similar features to the son. (figure 1) There were no anomalies in any siblings or near relatives. The patient was presented with normal intelligence and little speech difficulty. The enlarged size of the head was

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noticed by the mother when the boy was 9 months old and the severity had gradually increased.

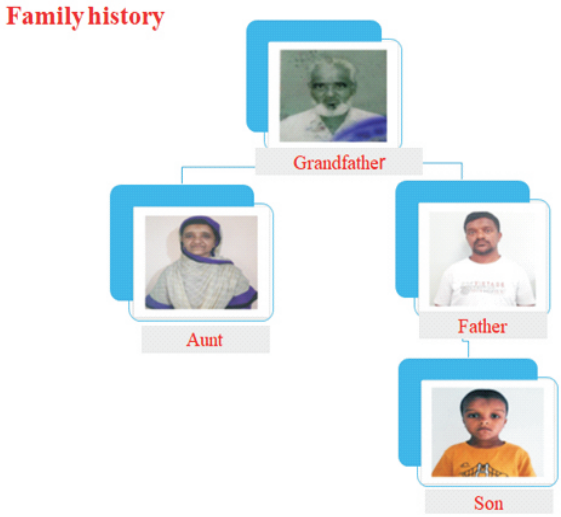


Figure1.Flowchartof detailed family history.

On examination:

On extraoral examination, the patient's head was dolichocephalic with enlarged cranial vault and frontal bossing, and the face was mesoprosopic, concave with short upper lips. Orbits were shallow with hypertelorism, bilateral proptosis, strabismus, depressed nasal bridge and hypoplastic maxilla with relative mandibular prognathism. (Figure 2). Also, his hands and feet photographs were taken which were normal in contrast to those with other severe craniosynostosis syndrome, where there is pronounced syndactyly of the extremities (Figure 3).

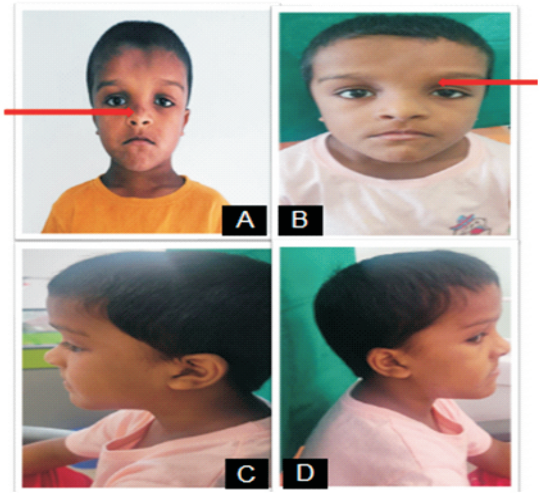


Figure2: Patient photograph shows (A) Parrot beak nose (Red arrow). (B) Hypertelorism and exophthalmos (Red arrow). (C) Side view shows hypoplastic maxilla (D) Prognathism of mandible.

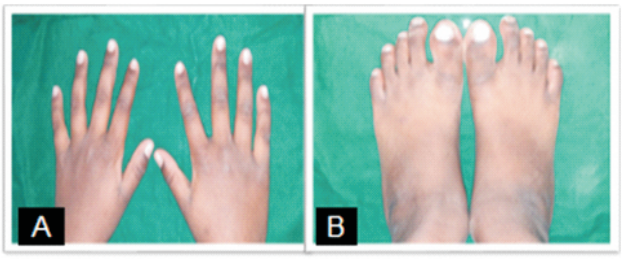


Figure 3: Photographs of hand and feet which were normal

On intraoral examination patient shows high-arched palate, V-shaped maxillary arch, wide spacing of upper teeth, bifid uvula, and anterior open bite. (Figure 4) Based on family history, intraoral and extraoral examination we can say that it might be a case of Crouzon's syndrome, to confirm it the patient was advised radiographic examination.

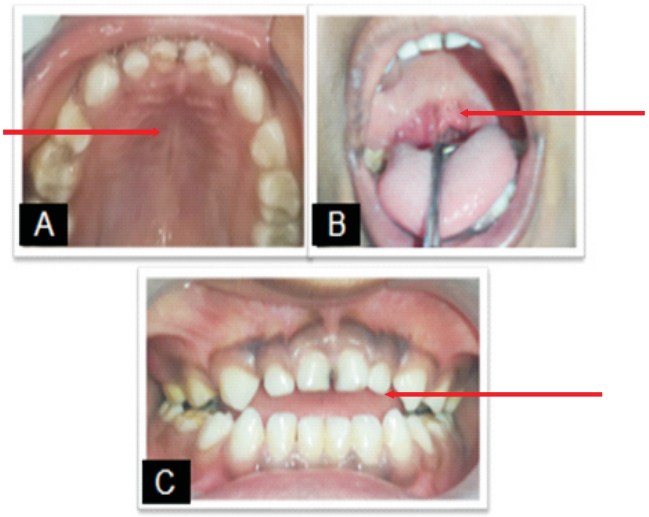


Figure 4: Intraoral examination shows (A) High arched palate (B) Bifid uvula (C) Anterior open bite

Radiographic examination:

The patient was subjected to radiographic investigations. The orthopantomogram revealed the presence of all deciduous teeth with narrow condyles and delayed eruption of teeth (Figure 5). The lateral view of the skull shows a discontinuity in the coronal border of the skull and copper-beaten appearance of the skull due to an increase in intracranial pressure (Figure 6). Hand wrist radiograph shows 5-carpel bones, out of which two shows slight ossification, radiolucency seen in two carpel bones, suggestive of a fracture line, and one metacarpal bone is not fused (Figure 7). The posteroanterior view of the skull shows multiple radiolucencies appearing as depressions resulting in a hammered or copper-beaten appearance due to increased intracranial pressure (Figure 8).

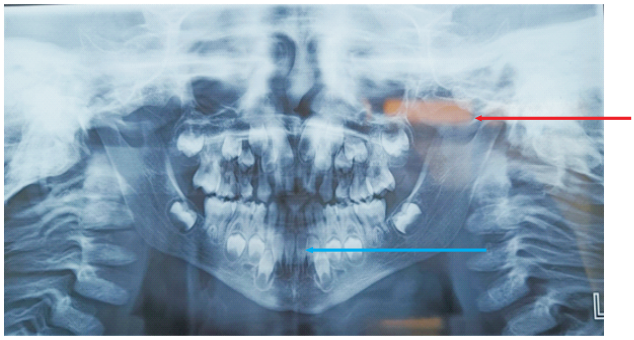


Figure 5: OPG shows narrow condyle (red arrow) and delayed eruption (Blue arrow).

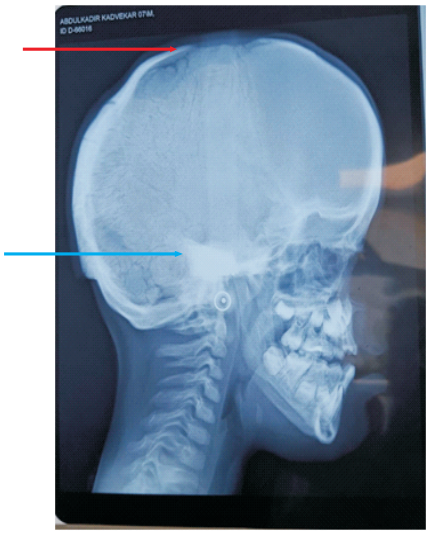


Figure 6: Lateral view of skull shows and discontinuity in coronal border of skull(red arrow) and copper beaten appearance of skull(blue arrow).

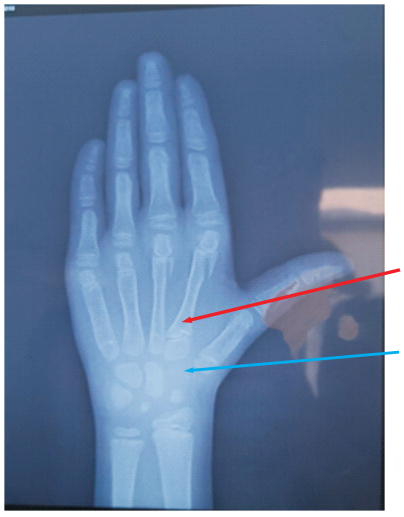


Figure 7: Hand wrist radiograph shows one metacarpel bone is not fused(red arrow) Two carpel bone shows slight ossification(blue arrow).

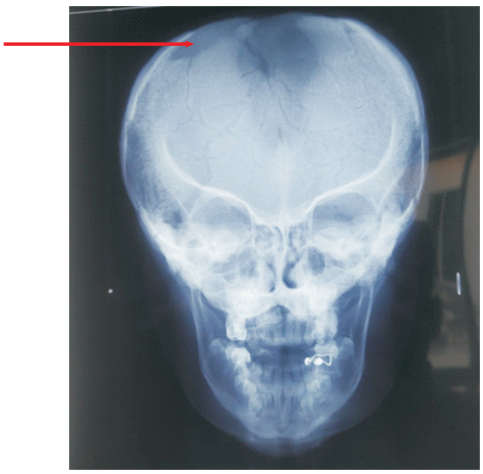


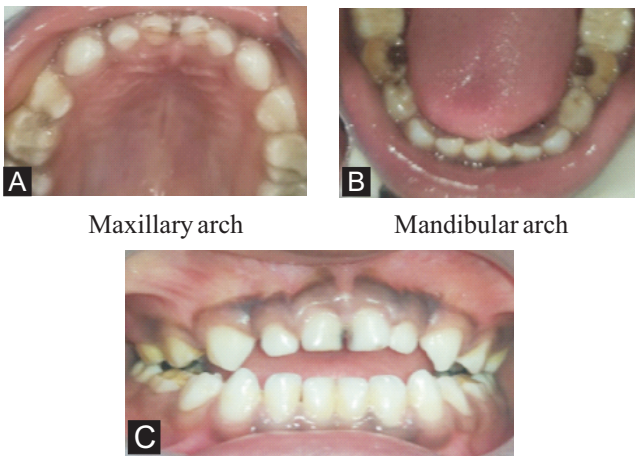
Figure 8: Posteroanterior view of skull shows copper beaten appearance of skull(red arrow).

The clinical, dental, ophthalmologic features and radiographic findings pointed to the diagnosis of **Crouzon syndrome**. Owing to the compromised oral hygiene, a thorough oral prophylaxis was performed.

Treatment plan:

Intraoral examination showed that the patient had early mixed dentition, with all primary teeth present in the maxillary and mandibular arch, in the maxillary arch deep proximal caries were present with 51, 55, 61 out of which 55 was pulpally involved. In the mandibular arch deep occlusal caries were present with 75, 85 and both were pulpally involved.(figure 9).

Intraoral Findings:



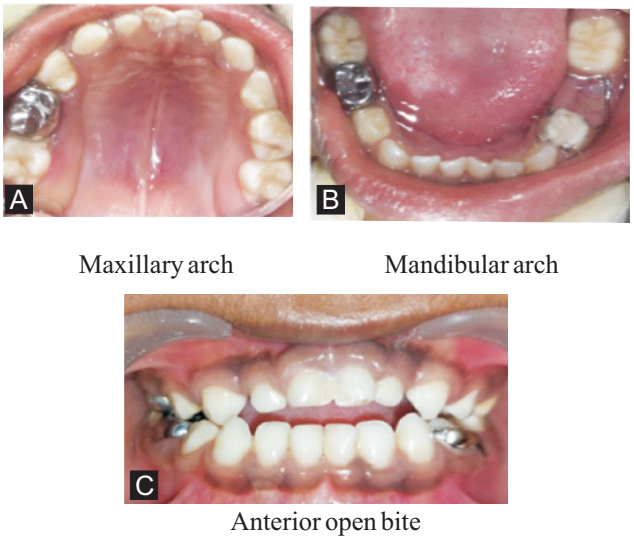
Maxillary arch Mandibular arch

Anterior open bite

Figure 9: Intraoral examination shows (A) Maxillary arch (B) Mandibular arch (C) Anterior open bite

In the first visit, full mouth ultrasonic scaling and polishing was done, pit and fissure sealant was applied on 16,26,36,46 using Tell-Show-Do technique and distracting the patient by showing movies during the procedure. While in the second visit, composite restoration was done for 51, 61. In subsequent visits patient cooperation was increased so pulpectomy was done for 55, 85 followed by stainless steel crowns. The extraction of the grossly decayed 75 was done and impressions of the maxillary and mandibular arches were made for the fabrication of a fixed space maintainer. Reverse band and loop space maintainer were delivered and postoperative photos were taken (figure 10). Orthodontic evaluation for the correction of maxillary hypoplasia, mandibular prognathism, and anterior open bite growth modulation therapy is planned with rapid maxillary expander(RME) and the face mask to enhance maxillary growth by expansion in the transverse plane, saggital advancement, and vertical lengthening thereby improving his facial profile and appearance.

Postoperative Photos



Maxillary arch

Mandibular arch

Anterior open bite

Figure 10: Postoperative photos shows (A) Maxillary arch (B) Mandibular arch (C) Anterior open bite

Discussion:

Octave Crouzon a French neurologist, in 1912, described the hereditary syndrome of craniofacial synostosis in a mother and son. He described the triad as skull deformities, facial anomalies and exophthalmos now known as Crouzon's syndrome.[5] The cause is attributable to mutations in the fibroblast growth factor receptor-2 (FGFR2)gene which is mapped to chromosome locus 10q25-10q26 but exhibits locus heterogeneity with causal mutations in FGFR-2 (Crouzon syndrome) and FGFR3 (Crouzon syndrome with Acanthosis nigricans) in different affected individuals.[6]

The most common ocular abnormalities are shallow orbits, ocular proptosis, orbital hypertelorism, strabismus, papilloedema, and visual loss.[7] There also have been rare occurrences of nystagmus, iris coloboma, blue sclera, glaucoma and luxation of the eye.[7] Headaches and seizures are attributable to elevated intracranial pressure. Conductive hearing loss is common owing to ear canal stenosis or atresia.[8] Upper airway obstruction develops secondary to septal deviation, mid-nasal abnormalities, nasopharyngeal narrowing and can lead to acute respiratory distress.[8]

In the mouth, hypoplastic maxilla, relative mandibular prognathism, short upper lip, malocclusions, and v-shaped maxillary dental arch have been reported, and also narrow, high, or cleft palate and bifid uvula.[9] Radiographs, Magnetic Resonance Imaging (MRI) scans, genetic testing, X-rays and CT scans can be used to confirm the diagnosis.[10] C.T scan brain shows signs of raised intracranial pressure, and fusion of coronal and sagittal sutures, and 3D images will reveal copper beaten appearance.[11]

The Management of Crouzon's disease is multidisciplinary and early diagnosis is important. Early craniectomy with frontal bone advancement is most often indicated to prevent or treat increased intracranial pressure.[2] If necessary, midfacial advancement and jaw surgery can be done to provide adequate orbital volume, reduce the exophthalmos, and correct the occlusion to an appropriate functional position.[10] Prognosis depends on the severity of malformations. Innovations in craniofacial surgery have enabled patients to achieve their full potential by maximizing their opportunities for intellectual growth, physical competence, and social interaction.[1]

Conclusion:

Crouzon's syndrome should be managed as early as possible as it results in poor cosmetic appearance and results in other complications like mental retardation, airway obstruction, and decreased visual acuity as the age advances. With proper treatment, these patients can be productive and active members of the mainstream society.

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