Prosthetic Rehabilitation of a Patient with Symptoms of Iron Deficiency Anaemia in association with Fraser's Syndrome – A Case Report

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Abstract

About a quarter of the population in the world is affected with Anaemia and deficiency of iron is one of the major causes. Associated symptoms are long term fatigue, cognitive function which shows impairment, and decreased well-being. "Fraser syndrome" can be described as rare, genetically inherited disorder. It is manifested as Crytophthalmos, Syndactyly along with Enamel Hypoplasia because of which the patient is referred to a dentist. The following report presents the report of a male patient who had symptoms of iron deficiency anaemia and also showed signs & symptoms of Fraser's Syndrome. Proper knowledge regarding Fraser's syndrome and iron deficiency anaemia is imperative to plan an oral rehabilitation treatment protocol for such patients.

Keywords: *Iron Deficiency Anaemia, Fraser's syndrome, Enamel Hypoplasia, Occular Prosthesis.*

Introduction

The main objective for the patients with Iron deficiency Anaemia is facilitation of the understanding and curing measures of the primary causative condition and avoidance of the treatment measures which are invasive and the risks with come along with it.[1]

The World Health Organization(WHO) has defined iron deficiency anaemia as the one which has haemoglobin values "Less than 7.7 mmol/l (13 g/dl) in men and 7.4 mmol/l (12 g/dl) in women". Iron deficiency anaemia is characterized by microcytic, hypochromic erythrocytes and depleted stores of iron in the body. The oral signs and symptoms include sensation of burning of the oral mucosa, lingual varicosity, xerostomia, Oral Lichen Planus, reduced sensory sensations of the oral mucosa and taste dysfunction.[2]

"Fraser syndrome" was first illustrated in the year of 1962 characterised by Syndactyly, Cryptophthalmos, oral clefting, genitor-urinary malformations, and laryngeal stenosis. The main cause is said to be mutations in "FRAS1", "FREM2", or "GRIP1" genes.[3]

The guidelines for diagnosis of the "Fraser's syndrome" is given by Thomas et al(1986) which states that the diagnosis meets in those patients which contains at least of two of the major criteria and minimum of one minor criterion or one of the major criterion and four of the minor criteria.[4]

Table No.1 Diagnostic Criteria for Fraser's Syndrome4

Major Criteria	1 Syndactyly. 2. Cryptophthalmos. 3 Affected Siblings. 4. Abnormal genitalia.
Minor Criteria	1 Congenital malformation of ears 2 Congenital malformation of nose 3 Cleft lip and/or palate 4. Congenital malformation of larynx 5 Umbilical hernia. 6. Skeletal defects 7 Mental retardation. 8. Renal agenesis

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Oral Manifestations for the syndrome include facial asymmetry, cleft palate along with conditions like Enamel Hypoplasia. Most common signs and symptoms which the patient presents are cryptophthalmos and, syndactyly. [5]

Case Report

A 24 year old Male patient reported to the private dental clinic with the chief complaint of missing teeth, caries and poor aesthetics. He was born second to the consanguineous parents .During his childhood he went multiple surgeries to correct genitalia abnormalities. The patient was born without one kidney and has not undergone any treatment for the same. His intellectual level was normal and no history of any medical disease or disorder was seen in the family.

Ophthalmologic examination showed cryptophthalmos with loss of vision. Head, Face and Neck check up demonstrated hypertelorism which was mild, slight facial-asymmetry and slight reduction in lower facial height.(Figure Nos.1)

Oral examination indicated microdontia which was generalized in nature and involved dentitions of arches with reduction in their size. He had poorly developed teeth with oral hygiene which was diagnosed with gingivitis and Hypoplasia of the enamel. Patient had reduced taste sensations and burning in oral mucosa which further hinted for diagnostic tests for Iron deficiency anaemia. Teeth present were 11-18, 21-28, 31-37 and 41-47 with grade II mobility seen with 11, 21,31, 41 and 42. Radiographic examination showed teeth with short roots (Figure No.2)

Management of Anaemia due to Iron Deficiency:

Blood tests were done to estimate the Haemoglobin level which revealed 11g/dl value. The peripheral smear showed hypochromic, microcytic, erythrocytes and low iron stores.(Figure No. 3)

Invasive tests like anti-parietal cell antibody, celiac serology, *Stool sample was tested for H. pylori infection* & faecal occult blood test were carried out. Iron replacement therapy was initiated to check the response for a span of 4 to 8 weeks. There was considerable improvement in the haemoglobin level of the patient (14g/dl) and follow up was carried out after 3 to 6 months.

Dental Rehabilitation:

To meet the requirements of the patient, a complete tooth supported overdenture was considered. A proper sequential treatment protocol was planned. Teeth with compromised periodontal support were extracted and Elective Endodontics was implemented in the remaining teeth and the preparation was done in a dome shaped contour which could receive copings.

Custom post patterns were fabricated and an impression was made. The copings were finished and polished and luted to the abutment teeth with the help of GIC luting cement (Figure No. 4).

After primary impression, border moulding was done and secondary impression was made. After Jaw Relation and a successful try in, processing was done using heat-cure acrylic resin. The denture was well accepted by the patient and had satisfactory results in terms of aesthetics and function.

Occular Rehabilitation:

An acrylic special tray with holes and a syringe tip was made for impression procedure[6] (Figure No. 5). Impression was made using light body elastomeric material. Indentations were marked for proper orientation of the cast which was obtained with respect to fabrication of further wax pattern. The wax pattern was fabricated so as to resemble and simulate the eye which was lost. The stock scleral eye was opted after comparing with the normal eye of the patient. That iris from the stock eye shell was embedded in the pattern. (Figure No. 6).

Flasking and Dewaxing was carried out for the adapted wax pattern. Packing was done with Self-cure tooth colour acrylic Polymerizing resin .After finishing and polishing spectacles were used to make the borders of the occular prosthesis inconspicuous. Necessary instructions for placement, cleaning, and prosthesis removal were given to the patient. Regular follow up appointments were scheduled (Figure No. 6).

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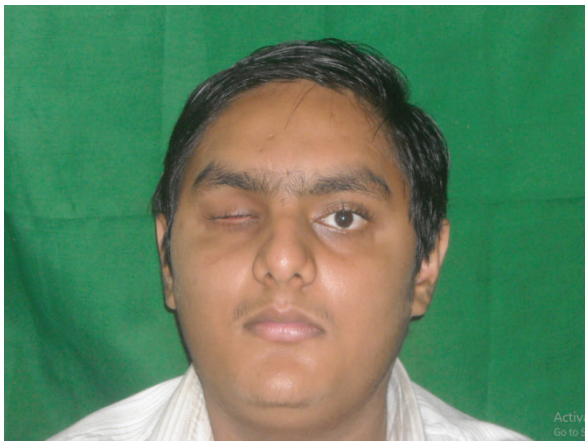
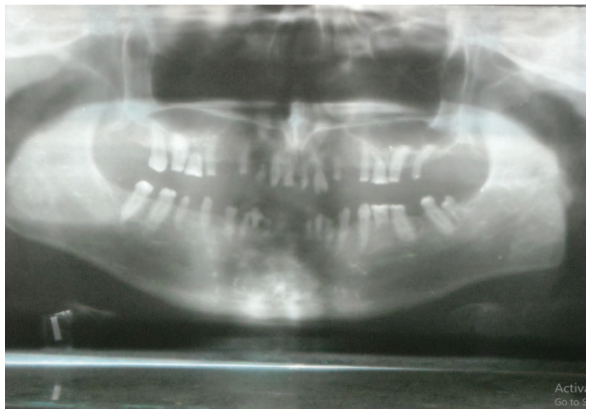
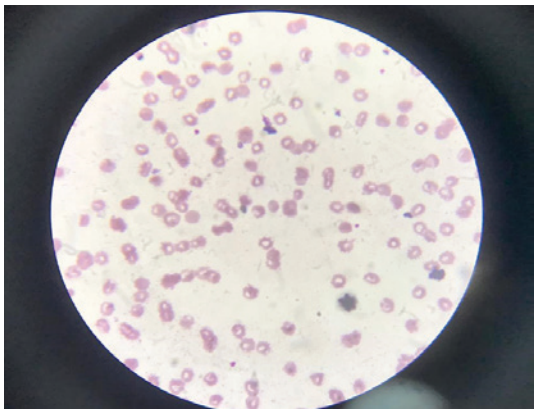
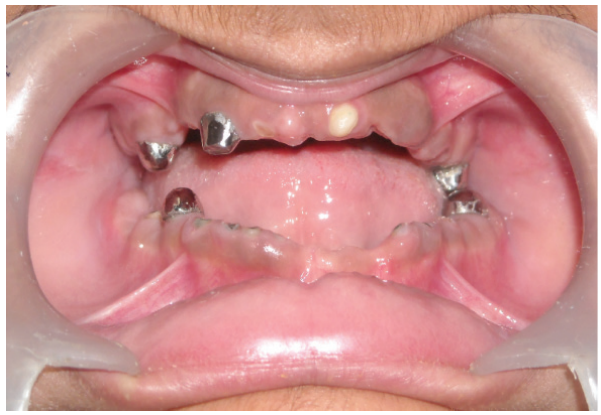




(Figure No. 1 Extraoral Examination of the Of the patient showing reduced lower Face height)

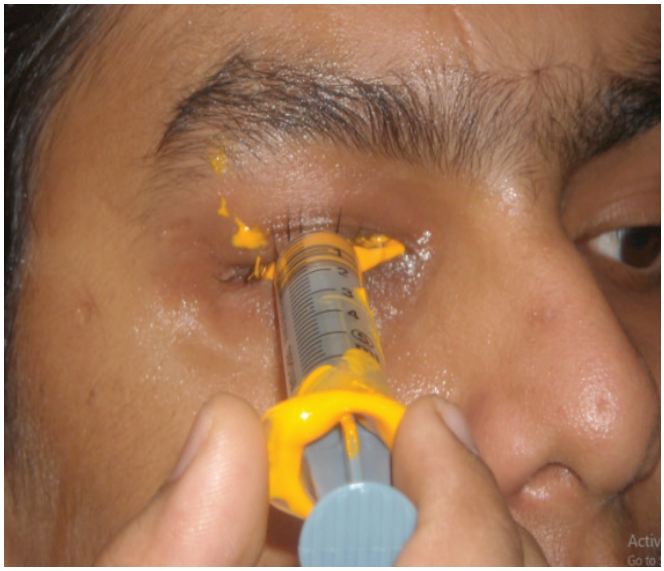
(Figure No. 2 Radiographic examination showed teeth with reduced face height)



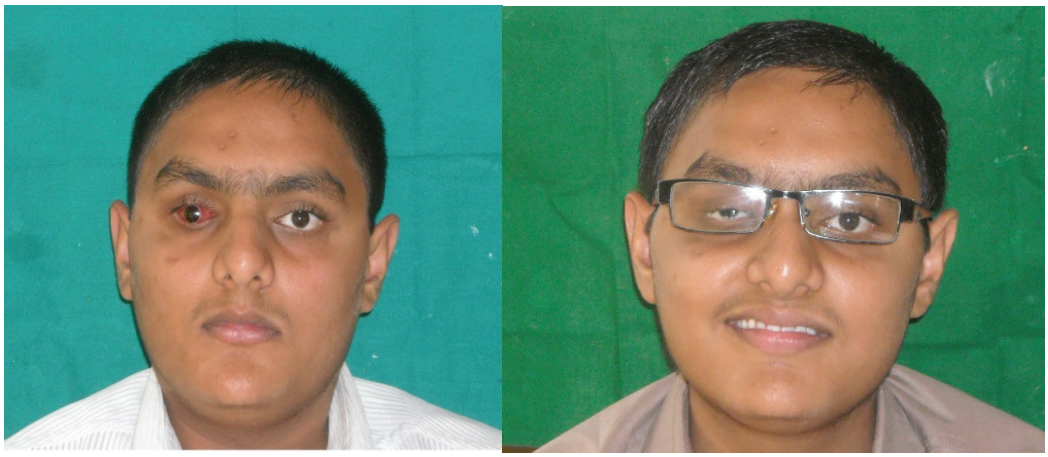


(Figure No.3 Peripheral Smear showing Microcytic, Hypochromic anaemia)

(Figure No. 4 The Metal copings Finished, Polished and Luted)



(Figure No.5 An acrylic special tray with holes and a syringe tip was made for impression procedure)



(Figure No.6 Try in of the occular Prosthesis and Final Prosthesis placed)

Discussion

There are several studies which state the importance of immediate and prompt care for the patients with anaemia associated with iron deficiency. It not only eradicates fatigue related symptoms but also elevated the level of quality of life. The treatment of iron deficiency anaemia associated with certain specific syndromes should be undertaken according to the guidelines of the specialist.

The oral supplemental therapy of iron deficiency anaemia may be limited to the patients which do not manifest gastrointestinal signs and symptoms such as cramps in the abdominal region and nausea along with vomiting. Intravenous iron therapy is another alternative for oral iron replacements that is indicated for the patients with blood loss more than 10 ml/ per day.

Fraser's Syndrome is an autosomal dominant but a rare disorder. Oral manifestations which were met by the patient were as described earlier which included short roots, retained deciduous teeth including Enamel Hypoplasia. [7] After diagnosis of anaemia caused by iron deficiency, identification of the cause should be done or else it will lead to recurrence even after proper treatment protocol.

Cardiovascular abnormalities which are seen in patients with Fraser's Syndrome include coarctation of aorta, valvular stenosis, ventricular septal defect atrial septal defect , and cardiomyopathies. If any involvement of cardiovascular system is seen, the protocol regarding infective endocarditis to decrease the microbial load should be carried out. When any sort of Renal Involvement is seen nephrotoxic drugs should be avoided.[7]

Conclusion

Anaemia due Iron deficiency is the major cause of fatigue and impairment in the cognitive function and diminishes the quality of life which is all the aspects of "Health status", "Lifestyle", "Life satisfaction", "Mental health" and "Well-being" reflecting together in an individual.[8] It is an important health outcome and hence Iron deficiency anaemia should be treated upon diagnosis.[9]

Proper knowledge of oral and dental features is definitely going to help with identifying the Fraser Syndrome. Dentistry should reconsider the recent aesthetic approach keeping in account of previous data working its way for successful prosthetic rehabilitation.[10]

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Conflict of Interest: None

Ethical Clearance : Obtained

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