Case report

Rehabilitation of a patient with chondroectodermal dysplasia

Srividya S*
Chandrasekharan Nair K.**
Jayakar Shetty***

Abstract

A 24 year old female patient reported with a complaint of missing upper and lower front teeth since childhood. Medical history revealed that the patient had preexisting cardiac condition, rheumatoid heart disease with mitral valve prolapsed and mild mitral regurgitation. General physical examination revealed, that the patient was of a short stature with moderate built and marked bilateral genu valgum. Intraoral examination revealed that the teeth were hypoplastic with high caries rate, resulting in presence of several large restorations in relation to 16, 17, 26, 27, 28, 37, 47 with 12, 22, 31, 32, 33, 34, 41, 42, 43, 46 missing. The diagnosis was a case of Ellis Van Crevelo Syndrome (chondroectodermal dysplasia.) Treatment plan involved oral prophylaxis, extraction of 83, restoration of carious teeth, frenectomy of hyperplastic low attachment labial frenum, replacement of missing teeth – mandibular anterior teeth with removable partial denture and maxillary fixed partial denture – 13 - 23. The management of this case involved a multidisciplinary team approach involving medical (cardiac surgeon, general physician) and dental team comprising of prosthodontist, endodontist, periodontist.

MeSH words: Chondroectodermal dysplasia, Ellis Van Crevel Syndrome

The most joyous moment in a parent’s life is the birth of a child. As the child grows and crosses its appropriate milestones of growth, it becomes a reason for celebration. However not all children in this world are so fortunate. There are many differently abled children in this world for whom taking a single step ahead is indeed a herculean task. These patients not only suffer from their physical disability but also have a psychological set back because of the social stigma attached with their disability. When a dental professional is called upon to provide service for such patients, it is important that we handle them with appropriate care, caution and patience. This case report is about a 24 year old female patient who complained of missing upper and lower anterior teeth since childhood. Her main concern was the appearance. The patient gave a history of first degree parental consanguinity with one younger sibling (sister) having similar condition. She had a son of 8 year old who has normal growth. It was evident that the patient was psychologically disturbed as well. General physical examination revealed that the patient was of short stature with moderate built (Fig1). Bimanual and bipedal hexadactyly was noted (extra digits on both hands and feet) (Fig 2,3). Closest examination revealed that the finger and toenails were markedly hypoplastic, incompletely formed and were small with longitudinal ridges. Nails were completely missing in relation to 5th and 6th digits of both right and left hands and feet. Vital signs were within normal parameters. Patient had pallor suggesting anemia. Extraoral examination revealed facial concavity due to mild maxillary hypoplasia, incompetent lips, protruding central incisors(Fig4). Intraoral examination revealed brownish black (melanin ?) pigmentation of buccal and labial...
mucosa, marginal and attached gingival and lateral borders of the tongue, markedly pronounced on the buccal mucosa along the line of occlusion. The labial sulcus of the upper and lower arches was crossed by multiple abnormal frenulii with high labial frenal attachment in relation to both upper and lower arches with presence of midline diastema in relation to upper arch (Fig 5). The erupted teeth were hypoplastic with high caries rate, resulting in several large restorations and root canal treatments in relation to 16, 17, 26, 27, 28, 37, 47 with 12, 22, 31, 32, 33, 34, 41,42,43,46 missing. 33, 34, 43, 46 were extracted due to caries and periodontal reasons two years ago(Fig 6,7). Angle's Class III molar relationship was noted on the left side. Wrist radiograph revealed the presence of an extra digit in the ulnar side polymetacarpalism, syncarpalism, shortening of middle and distal phalanges (Fig 8). Radiographs of lower limbs revealed hypoplasia of proximal tibia epiphysis with mesial angulation and shortening of tibia and fibula of both the limbs. Orthopantomograph revealed the presence of retained 83, impacted 35, and missing 12,22,31,32,41,42 and 46 with hypoplasia of left condyle (Fig 9). Both extraoral and intraoral findings and radiographic investigations suggested a case of