CASE REPORT

TUBERCULAR INTRANASAL MESIODENS IN ORO-FACIAL-DIGITAL SYNDROME

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ABSTRACT

Ectopic eruption of teeth has been reported in mandibular condyle, coronoid process, orbit, maxillary antrum, palate, nasal septum and chin. But an erupted supernumerary nasal tooth is rare. Although asymptomatic in some, a nasal tooth has the potential to cause epistaxis, septal perforation, rhinorrhea and several other complications. It is still rare for a nasal tooth to be associated with Oro-facial-digital syndrome which is a genetic malformation involving oral, facial and digital structures accompanied with varying degrees of mental retardation. The oral and dental manifestations of this syndrome include hypertrophic frenula, lingual hamartomas, ankyloglossia, dental caries, anomalous anterior teeth, enamel hypoplasia, supernumerary teeth and missing teeth. Presented is a case of an erupted tubercular mesiodens in the nasal cavity of a boy aged ten years. To the best of our knowledge, this is the first reported case of a nasal tooth in a child with oro-facial-digital syndrome. Children born with structural anomalies of the face, oral cavity and digits should be thoroughly evaluated to rule out the possibility of syndromes. Moreover, a multidisciplinary approach is required to render the best treatment to the patient. Also, a nasal tooth should be considered a possibility amongst undiagnosed nasal masses.

Key words: *Ectopic eruption, nasal tooth, oro-facial-digital syndrome, supernumerary tooth*

INTRODUCTION

A tooth in the nasal cavity was initially reported in 1897 with an age range of 3 to 62 years.[1] Approximately 78 cases of intranasal teeth have been reported.[2] A literature survey from 1959 to 2008 identified only 25 supernumerary nasal teeth in 23 patients.[3] The prevalence of an erupted intranasal tooth is 0.61% in bilateral cleft lip and palate and 0.40% in unilateral cases.[4]

The term 'Oro-facial-digital dystosis' includes a triad of malformed oral, facial and digital structures.[5] The incidence is 1:2,50,000 births and 1:100 in patients with cleft palate. A total of thirteen variants have been listed.[6]

CASE HISTORY

A ten-year old boy wanted a hard white tooth-like structure in the left nostril removed; citing aesthetic reasons (Fig 1). His left cleft lip and alveolus was repaired when he was one year old. He was congenitally deaf and dumb with delayed developmental milestones. His mental age was ten years on Development Screening Test and his performance Intelligence Quotient on Seguin Form Board Test 125, revealing an above average intelligence. The family history was non-conclusive.

General examination revealed ocular hypertelorism, depression of nasal bridge (Fig 2), syndactyly of

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both hands and feet along with unilateral duplication of the hallux or the great toe (Figs 3, 4). There was also an associated bilateral conductive deafness along with speech disorder. However, cardiovascular, pulmonary system and his biochemical parameters were within normal limits. An intra-nasal examination showed a hard immovable mass, resembling a tooth crown, just at the entrance of the left nostril at the scar terminus. There was no oro-nasal communication.

An intra-oral examination showed retained primary mandibular central incisors 71, 81 (FDI notation). A disto-labial rotation of permanent maxillary left central incisor 21 and hypoplasia of permanent maxillary left lateral incisor 22 was observed. Signs of cleft alveolus were also observed on the left (Fig 5).

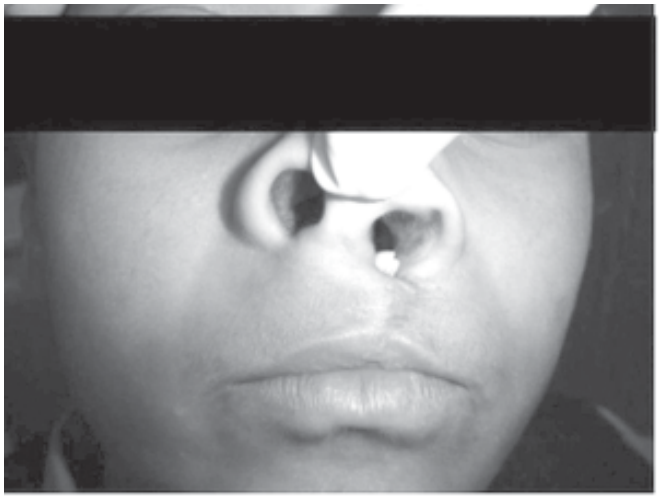

Fig 1: Tooth in left nostril with scar of repaired cleft lip

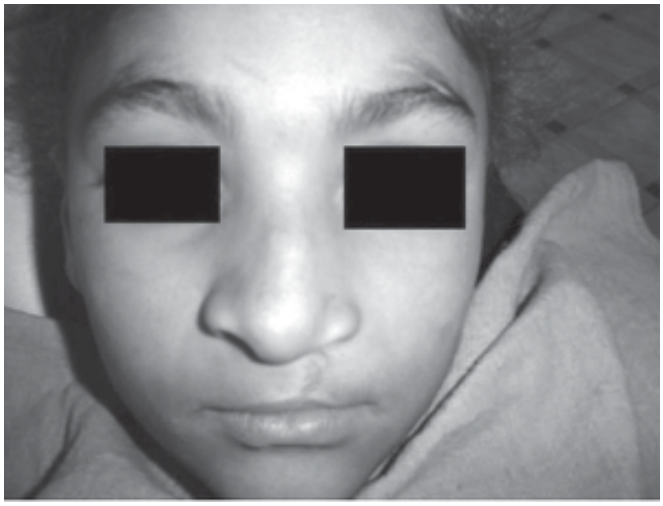

Fig 2: Hypertelorism, depressed nasal bridge and cleft lip repair

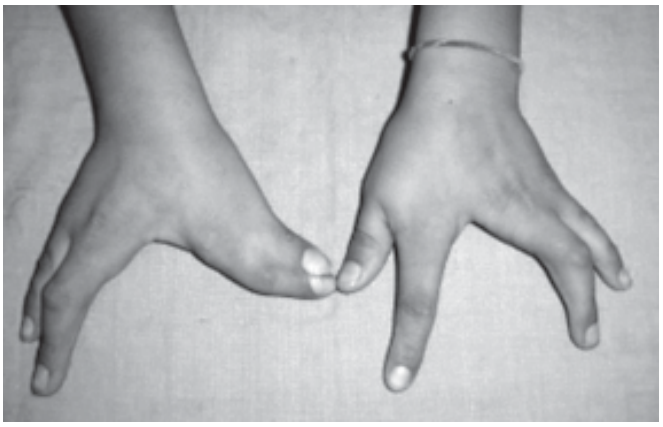

Fig 3: Syndactyly of hand

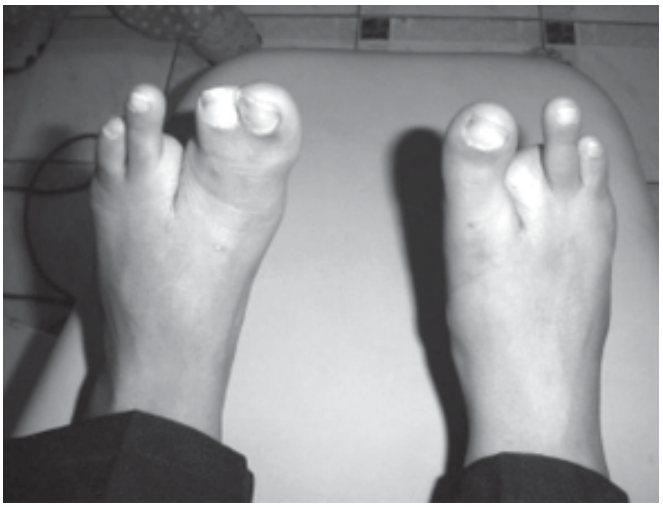

Fig 4: Syndactyly of feet with unilateral duplication of hallux

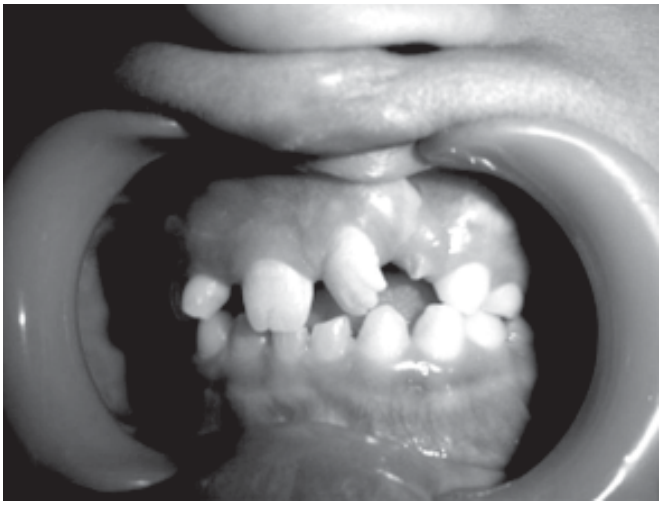

Fig 5: Cleft alveolus, rotated 21, hypoplastic 22, retained 71, 81

Radiographic examination revealed a single-rooted, dilacerated intranasal supernumerary tooth with agenesis of permanent mandibular central incisors (Figs 6, 7).

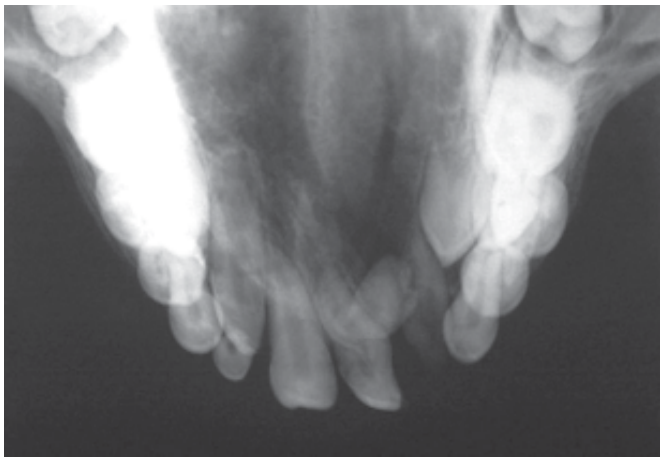

Fig 6: Maxillary occlusal radiograph showing cleft and the nasal tooth

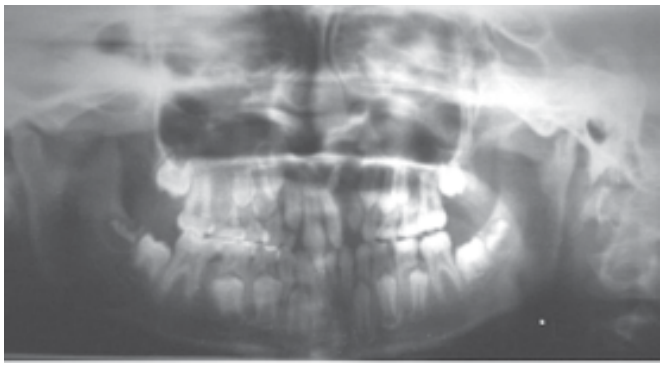

Fig 7: An OPG showing the nasal tooth, cleft and agenesis of 31, 41

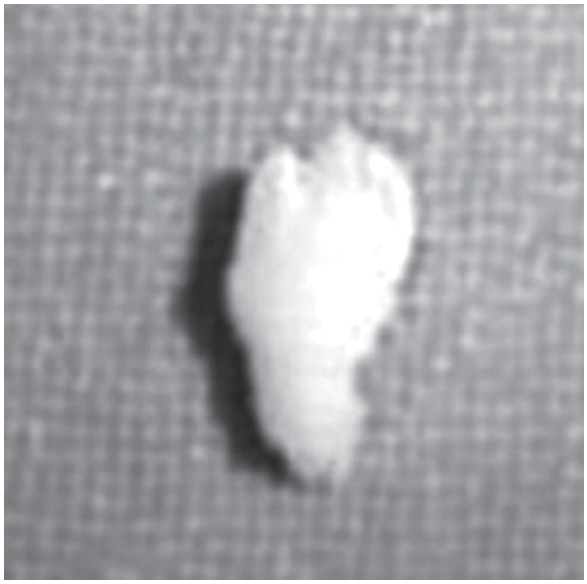

Fig 8: Extracted supernumerary nasal tooth

The nasal tooth was extracted under local anesthesia. It was tubercular in shape with mammelons (Fig 8). A nasal pack arrested bleeding and post-operative healing was uneventful. The mother acted as an interpreter for the child and sign language was used throughout the procedure.

DISCUSSION

Ectopically erupted teeth have been detected in aberrant locations such as anterior mediastinum, retroperitoneal area, presacral area, coccygeal area, testes and ovaries. Although reported in mandibular condyle, coronoid process, orbit, maxillary antrum, palate, nasal septum and chin, yet a nasal tooth is rare.[1] Endicott in 1934 reported the first nasal tooth in association with cleft lip and palate.

Possible etiological factors of a nasal tooth are trauma, infections, obstruction to eruption, retained primary teeth, genetics, syndrome and cleft lip and palate.[1,4,7] In this case likely etiology of nasal tooth could be improper fusion of embryonic processes resulting in a cleft palate with a possible displacement of the tooth bud in the intranasal area. Moreover, trauma sustained during the surgical repair of the cleft alveolus during the first year of life could also be an additional etiological factor.

Clinically, a nasal tooth is frequently found on the floor of the nasal cavity. It appears as a hard white mass, often surrounded by granulation tissue and debris. The differential diagnosis includes a foreign body, rhinolith, inflammatory lesion with calcification secondary to syphilis, tuberculosis or fungal infection, bony sequestra, neoplasm, exotosis and odontoma.[7,8] Diagnosis is usually made on clinical and/or radiographic examination.

Signs and symptoms associated with nasal tooth include nasal obstruction, nasal discharge, abscess, ulceration, recurrent epistaxis, rhinitis caseosa, oronasal fistula, chronic sinusitis, facial pain, headaches, external nasal deviation, speech problems and occasionally no symptoms.[1] Aspergillosis,[8] lacrimal duct obstruction, rhinosinusitis, nasal septal perforation,[7] oronasal communication and osteomyelitis are varying degrees of morbidity associated with a nasal tooth. In the present case, the patient was asymptomatic and it was primarily due to esthetic reasons that he had sought dental intervention.

An asymptomatic intra-nasal tooth may be retained with periodic radiographs and clinical follow-up.

However, because of the potential to cause severe complications, it is advisable to extract the nasal tooth either via transpalatal or transnasal route. Recently, endoscopic extraction has been advocated for accurate dissection and better visibility.[1]

Hypoplastic teeth have also been reported in repaired cleft lip and palate with a high frequency of 92% in permanent maxillary anteriors.[9] A multifactorial theory highlights the role of genetics, postnatal environment, nutrition and surgical influences. In the present case, etiology of hypoplastic tooth 22 could be the cleft itself, trauma induced during surgical repair coupled with nutritional deficiencies.

Oro-facial–digital syndrome includes oro-dental anomalies with cleft lip and palate quite commonly found.[10] A midline pseudocleft in the inferior vermilion border of the upper lip is more common, although asymmetric true clefts can occur. Other facial/oral manifestations are ocular hypertelorism, aplasia of alar cartilage and alopecia, hypertrophic frenula, lingual hamartomas and ankyloglossia, dental caries, enamel hypoplasia, supernumerary and missing teeth. The digital manifestations are brachydactyly, polydactyly, syndactyly, clinodactyly and duplicated hallux. Some may have brain malformation and mental retardation, alteration of stapes and polystic renal disease.[6,10] The present case had ocular hypertelorism, repaired cleft lip and alveolus of left side, enamel hypoplasia, agenesis of permanent mandibular central incisors, supernumerary nasal tooth, syndactyly of both hands and feet with unilateral duplication of hallux and associated conductive deafness with speech disorder.

The management of oro-facial-digital syndrome is multidisciplinary. Cosmetic surgery for cleft lip and palate, tongue nodule and accessory frenula is usually required. Orthopedic surgery is often recommended to repair the defects of digits. Reconstructive surgery of auditory ossicles may be required to improve conductive deafness. The degree of learning disabilities and other cognitive impairment should be evaluated along with speech therapy to provide appropriate support. The dental treatment involves extraction of nasal tooth followed by orthodontic alignment and prosthetic rehabilitation.

CONCLUSIONS

It is important for medical specialists to include nasal teeth in differential diagnosis of nasal masses. Complications can and do arise with a nasal tooth but co-existence with syndromes should also be ruled out.

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