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Research Article

A CASE OF ACKERMAN'S SYNDROME: DISCOVERED BY SERENDIPITY

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ABSTRACT

Background: Ackerman's syndrome is the rarest of the rare syndromes with a documented prevalence rate of less than one in a million cases[1]. This syndrome has been known to present with Pyramidal molars, abnormal upper lip, wide philtrum, clinodactyly or syndactyly and sometimes also associated with juvenile glaucoma. Pyramidal molars are essentially single rooted molars, a rare morphological variation from the usual multi rooted molars and have very rarely been reported in the deciduous dentition. It is said to occur due to the failure of invagination of the Hertwig's epithelial root sheath.

Case report: Patient had reported to the department and a detailed examination and radiographic co-relation revealed the possibility of Ackerman's syndrome.

Conclusion: This is a report of a case of Ackerman's syndrome with pyramidal first deciduous molar in an eight year old female patient diagnosed by chance.

INTRODUCTION

Ackerman's syndrome is a documented familial syndrome inherited as an autosomal recessive trait and found to present with pyramidal molars/ taurodontism, sparse body hair, abnormal upper lip, wide philtrum, clinodactyly or syndactyly, increased pigmentation of finger joints and sometimes juvenile glaucoma.[2]Molars are multi-rooted. Usually, maxillary molars have three roots and mandibular molars have two roots. But, various types of morphological anomalies have been reported till date. Of which a rare finding is aPyramidal molar. Deep taurodontism or fusion of roots could also give rise to a single root. Variations in root forms have been reported to resultfrom failure of invagination of Hertwig's epithelial root sheath. This is because root growth depends on a continuous elongation of HERS.It has been demonstrated that root dysmorphology arises from failure of invagination of HERS.[9] This is a case report of Ackerman's syndrome in an eight year old female patientidentified by serendipity.

CASE REPORT

An eight year old female patientreported to the department with the chief complaint of spontaneous pain and sensitivity in the lower right posterior region of the jaw since the past few days, which aggravated in supine posture. A detailed history revealed multiple extractions of decayed teeth at a rural health care centrein the past. A detailed family history revealed that the patient was the only child of parents of consanguineous marriage. Clinical examination revealedattrition with 84, with adistorted morphology resembling 83 (i.e.; a deciduous canine). It was also noted that 74, 75 and 85 were prematurely extracted due to caries. There was generalised hypoplasia present. To rule out any anomaly of the permanent successors and to subsequently select the best line of treatment an orthopantomogram was adviced. Hypodontia in patients with Taurodontism have been reported earlier.[4] The orthopantomogram (figure 4) revealed that 84 (deciduous first molar) had a conical root in addition to a canine like coronal structure. Taking into consideration the overlying bone height of the permanent successor, 44 and that the tooth of interest 84 had chronic irreversible pulpitis;single sitting pulpectomy was selected as the line of treatment to retain the tooth till physiological exfoliation.Pulpectomy should be considered as an alternative to extraction in the cases of nonvital or irreversibly inflamed primary teeth.[5]Zinc oxide eugenol obturation was avoided as its potential of interferences with normal exfoliation of the tooth exists.[11]Optimal obturation was done using Metapex (Meta Biomed, Korea). It was established pre, per and post operatively that the 84 (FDI notation) was a pyramidal deciduous molar.

Other extra-oral and intra-oral findings included broad/wide philtrum as seen in (Fig 2), asymptomatic macular lesions (Fig 5) on the junction of hard and soft palate. A retrognathic mandible as seen in the Lateral Cephalogram (Fig 3) could be attributed to the early extraction of mandibular deciduous molars.Also, Clinodactyly of the left forearm, 5th digit was seen as in (Fig 6)

Illustrations

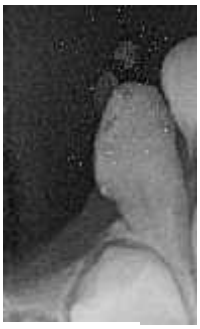

Figure 1

Figure 2

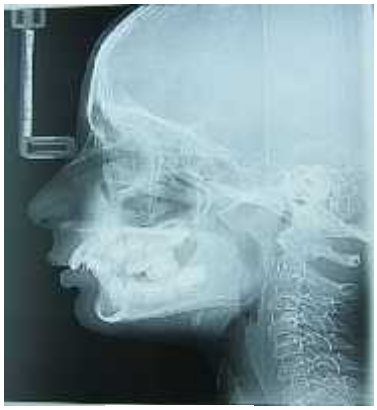

Figure 3

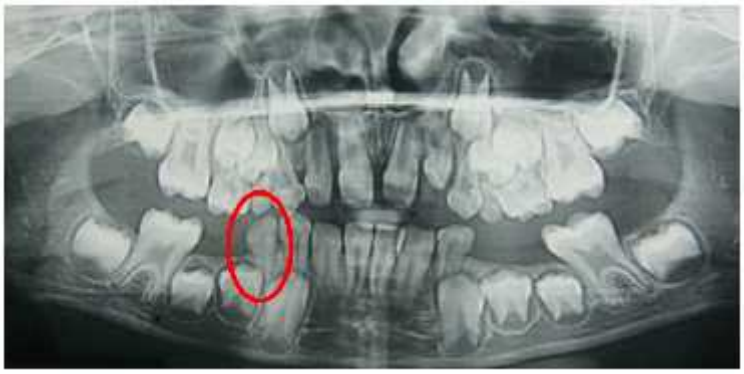

Figure 4

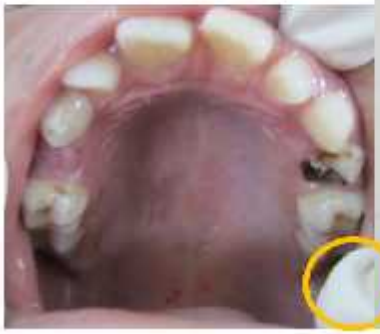

Figure 5

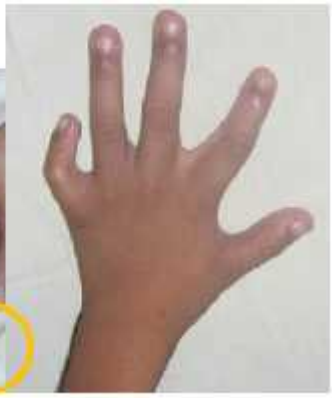

Figure 6

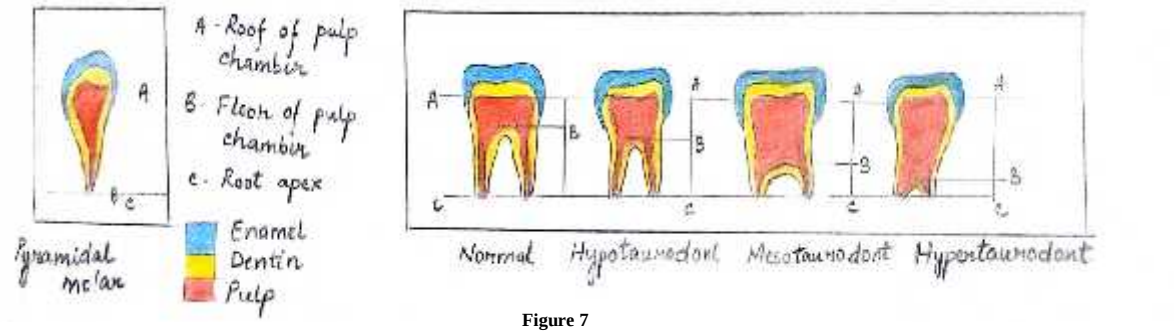

Figure 7

1. Intra oral periapical radiograph
2. Extra oral photograph showing broad philtrum
3. Lateral Cephalogram
4. Orthopantomogram
5. Intraoral photograph showing macular lesions
6. Clinodactyly of 5th finger of left hand
7. Schematic representation of pyramidal molar and different types of taurodontism

DISCUSSION

In a kindred study consisting of 20 members of a family of English-German descent with no consanguinity,radiographs of the molar teeth had revealed pyramidal molars and a co-relation with the other clinical findings like abnormal upper lip with broad philtrum, clinodactyly or syndactyly that were seen in all subjects of whom two siblings were identified with juvenile glaucoma for the firsttime documented and established Ackerman's syndrome (Ackerman 1973).[3] An anomaly of a single root has been appertained to fusion of roots or deep taurodontism. The first case of single rooted primary molars in a 10 year old child was reported by Ackerman *et al* and the next case was reported two decades later(Gideon Holan *et al*, 1991).[6] Ever since there have been just a handful of documented cases of deciduous pyramidal molars l*et al*one Ackerman's syndrome.

There lies a thin line of demarcation between a pyramidal tooth (with conical/tapering root) and hyper taurodontism (Fig 7). Taurodontism is a rare dental anomaly of multi-rooted teeth which is characterized by a vertically elongated pulp chamber and apically displaced pulpal floor.[7] Thus, to differentiate the two by a routine radiographic technique alone could be difficult and requires good pre and per-operative clinical co-relation.

Taurodontism is hereditary and of various types (Fig 7). Taurodont, pyramidal and fused roots may be aberrations of a single heritable trait.Confluent roots with separated pulp canals have been termed as fused roots, but the pyramidal teeth refers to teeth with single and wide root canal and has been reported as an autosomal dominant trait.[9]

Though three dimensional radiographic techniques are being used widely for determining morphological anomalies, their use in this case was not found to be mandatory. Since, the routine radiographic techniques were found to suffice in this case. Although three dimensional imaging techniques like Cone beam CT (CBCT), computed tomography (CT), spiral (SCT) or helical CT can be used for accurate evaluation of root canal morphology, using routine radiographs is preferred in young children.[10]

There is always a possibility of confusion between pyramidal molar, hyper taurodontism and a supernumerary tooth. Initially the tooth of interest, 84 was believed to have hyper taurodontism, but clinical inspection of the pulpal floor and radiographic co-relation confirmed a single rooted tooth with a solitary enlarged canal, which has been referred to as pyramidal molar. The easiest method to rule out supernumerary tooth is to check clinically and co-relating with an orthopantomogram.

CONCLUSION

The patient presented with a pyramidal molar, wide philtrum, clinodactyly of 5th finger of left hand and generalised hypotrichosis which confirmed it as a case of Ackerman's syndrome.

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