DOI: 10.26717/BJSTR.2018.05.001148
Nusrat Nazir. Biomed J Sci & Tech Res

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574-1241

Short Communication Open Access

Big things in Little Ones: an Original Study

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Received: May 15, 2018; **Published:** May 31, 2018

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Introduction

Pediatric dentistry is an age-defined specialty that provides both primary and comprehensive preventive and therapeutic oral health care for infants and children through adolescence, including those with special health care needs. Pediatric care is actually a multidisciplinary treatment approach which include;

a. Pediatric dentists.

b. General dental practitioners and other dental specialists.

c. Physicians and other health care providers.

d. Government agencies and health care policy makers.

e. Individuals interested in the oral health of children [1].

Rare diseases are a group of more than 5000 diseases that account for 10% of human disease [2-4]. In the last decades the attention of several disciplines of medicine have been focused on rare diseases; rare diseases are estimated to be 5000, and every week 5 new rare diseases are considered as new findings hence the number of these diseases is enormously increasing. In this articles we will be discussing all the rare cases and some less common cases that were referred to our department for evaluation and diagnosis.

Methodology

This prospective cohart study was conducted in the department of oral medicine and radiology government dental college Srinagar. Pediatric patients with some rare diseases /syndromes or complicated conditions were selected from the outpatient clinic over a period of 1 year [2016]. A detailed history and examination were taken with emphasis on chief complaint and its duration, general health of the patient, past medical history, and oral hygiene status. A thorough clinical examination was carried out in all patients pertaining to the type of disorder they were suffering. Appropriate investigations were done in all the cases to reach to a final diagnosis.

Statistical Analysis

Statistical software SPSS (version 20.0) and Microsoft Excel were used to carry out the statistical analysis of data. Data was presented by means of bar charts and the pie diagrams.

Results

A total of 35 patients with some special findings were included in the study. There were 17 males and 18 females. Following cases were seen during the period of one year (Table 1).

Table 1.

Ectodermal dysplasia	4
Ichthyosis	1
Firomatosis gingivae	3
pleomorphic adenoma	4
hypoplasictic condyle	1
epidermolysis bulosa	1
cleft lip+palate	5
cleft palate	6
amelogenesis imperfect	3
dentinogenesis imperfecta	2
lymphangioma tongue	1
selective Ig A deficiency	1

Discussion

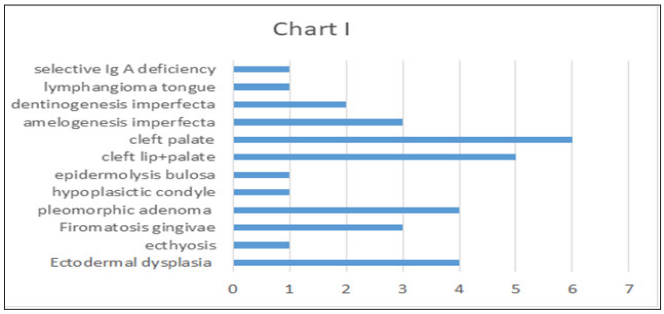


Figure 1.

In the discussion each condition is briefly described followed by the pictures of the cases. Pleomorphic adenoma is called as "mixed tumor" because it has both epithelial and connective tissue elements. It commonly affects parotid gland followed by minor salivary glands. About 450-750 minor salivary glands are present

in the head and neck region. Highest concentration of these minor salivary glands has been described on the palate, particularly the junction of hard and soft palate. Most common minor salivary gland tumors occur at this site [5]. In all 8-10% of salivary gland tumors arise in the palate. These tumors are malignant in 40-82%of cases [6-9]. There were 4 case of pleomorphic adenoma which came to our department diagnosed by FNAC and USG. Three were present on palate and one in submandibular region (Figures 1-4).

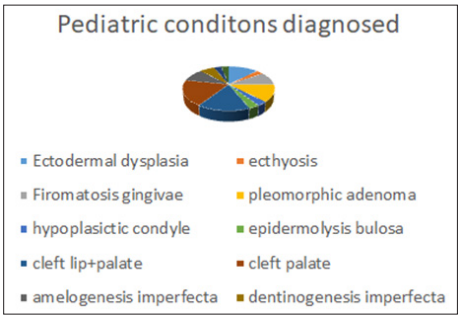


Figure 2.

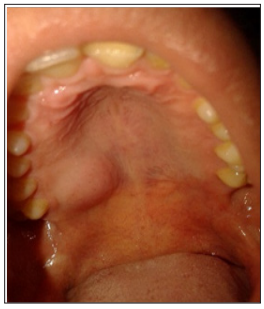


Figure 3.

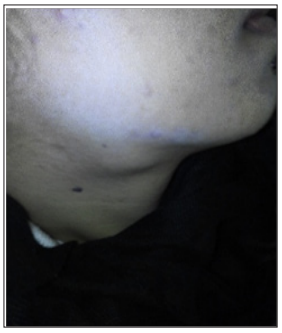


Figure 4.

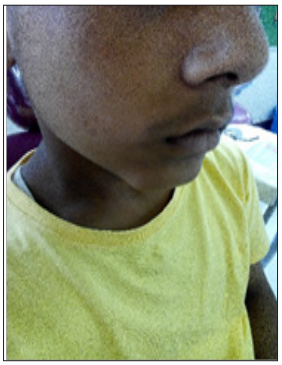


Figure 5.

Fibrous dysplasia (FD) is a bone development anomaly characterized by hamartoma proliferation of fibrous tissue within the medullary bone, with secondary bony metaplasia, producing immature, newly formed and weakly calcified bone, without maturation of the osteoblast which appears radiolucent on radiographs, with the classically described ground-glass appearance [10]. One such case was seen with involvement of mandible Figure 5 showing enlargement of chin and Figure 6 showing CBCT image of same patient.

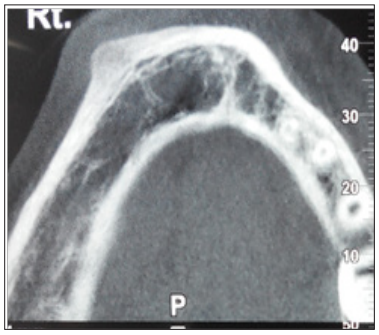


Figure 6.

Epidermolysis Bullosa (EB) is a group of rare inherited disorders, usually detected at birth or early childhood. This dermatological condition is a severe autoimmune disease [11-12]. Scarring of the extensor surfaces of the extremities, hands and feet are typical; milia occur frequently; and nails often become thick and dystrophic or are lost. The disorder affects both sexes equally and occurs in all racial and ethnic groups [13]. Figure 7 showing crustation of lips and Figure 8 is showing severe skin involvement and missing nails. Ichthyosis forms clinically and etiologically a large heterogeneous group of cornification disorders that are characterized by accumulation of hyperkeratotic scales on the skin surface [14,15]. Ichthyosis is derived from the Greek word "ikhthus" meaning "fish" and refers to the similarity in appearance of skin to fish scales. Ichthyosis is caused by abnormality in keratinization and exfoliation of the horny cell layer [16]. The relevant clinical feature of this disorder is scaling and thickening of cornified layer which is often accompanied by inflammation of the skin presenting itself as erythroderma [17]. (Figures 9-11) is showing the involvement of face legs and arms. There was genralised cornification of whole body in this patient.

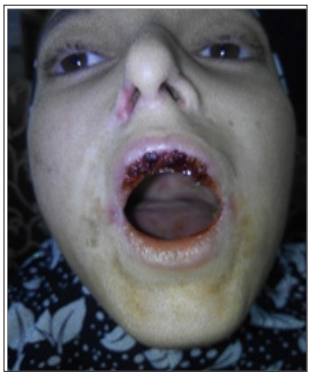
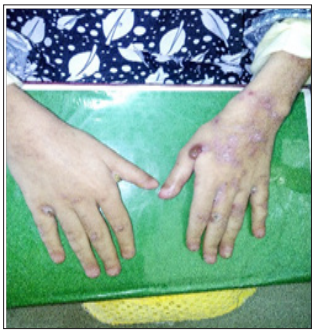


Figure 7.



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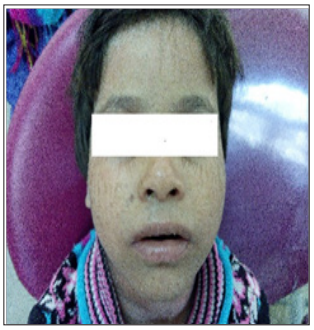

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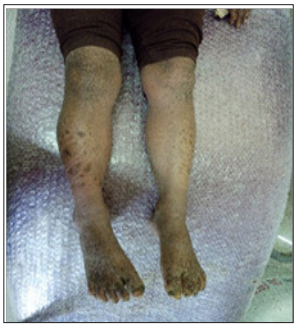

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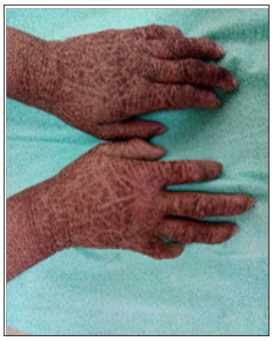
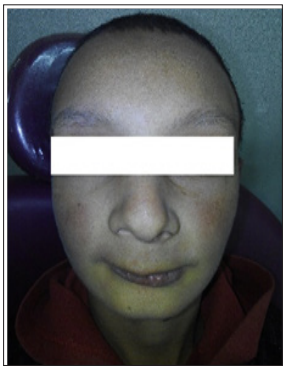


Figure 11.

Ectodermal dysplasia, as first described by Thurman, is a hereditary disorder occurring as a consequence of disturbances in the ectoderm of the developing embryo. The triad of nail dystrophy (onchodysplasia), alopecia or hypotrichosis (scanty, fine light hair on the scalp and eyebrows), and palmoplantar hyperkeratosis is usually accompanied by a lack of sweat glands (hypohidrosis) and a partial or complete absence of primary and/or permanent dentition [18-21]. (Figures 12-14) is showing hypotrichosis, onchodysplasia and oligodontia respectively. (Figures 15-18) are the cases of cleft lip, cleft palate, lymphangioma of tongue and fibromatosis gingivae respectively.



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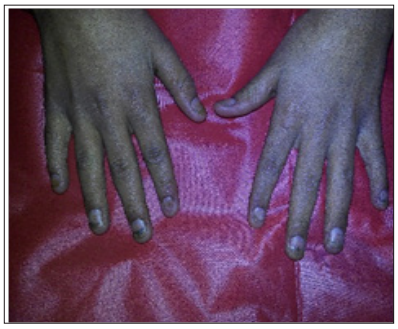


Figure 13.

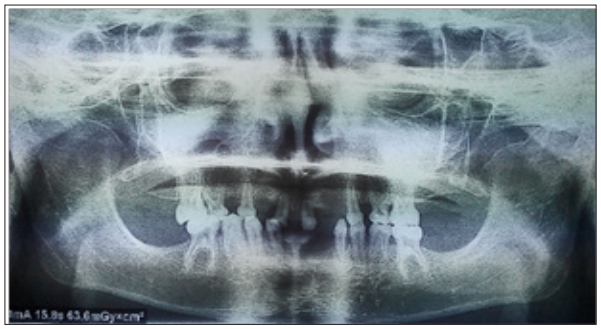
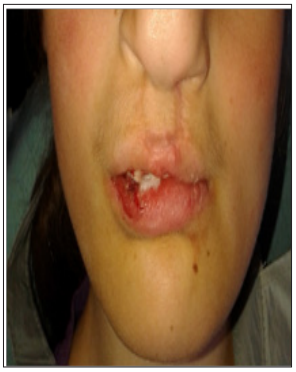


Figure 14.



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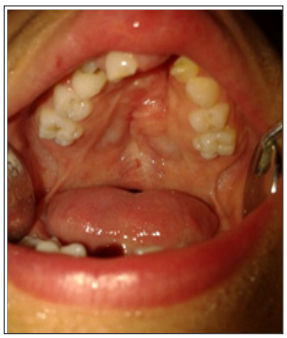

Figure 16.

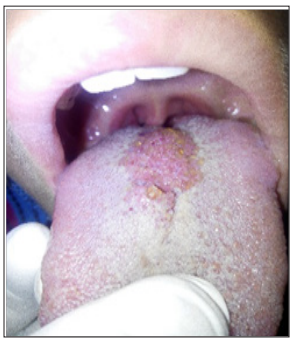

Figure 17.

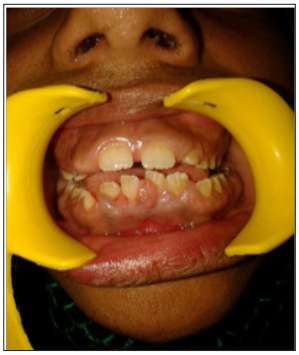

Figure 18.

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