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COMPARISON OF ORAL, CRANIOFACIAL AND RADIOGRAPHIC FEATURES IN NOONAN AND TURNER SYNDROMES

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ABSTRACT

Noonan syndrome and Turner syndrome are characterized by similar phenotypes, mainly, short stature, low birth weight, triangular face, prominent ears and webbed neck. This study clinically and radiographically documented and compared the oro-dental and craniofacial manifestations in both syndromes to assess the important differences and similarities that might be of diagnostic value. Three groups were included for comparison, group I consisted of 10 normal control females, group II consisted of 10 cases of Noonan syndrome, group III consisted of 10 cases of Turner syndrome. All cases were subjected to pedigree analysis, clinical examination, cytogenetic findings, clinical photographs, lateral and postero-anterior cephalograms and panoramic radiographs. The results were subjected to statistical analysis of variance (ANOVA) which showed that the similarities in both syndromes were high arched palate, thick lips, crown and root formation abnormalities, malocclusion, mandibular retrognathia, shortened posterior cranial base length and both maxillary length and width. Apparent hypertelorism was due to soft tissue telecanthus not a true bony hypertelorism. Noonan syndrome differed in the presence of tongue anomalies, increased mandibular downward tipping and vertical jaw dimension; shortened anterior cranial base length, mandibular width and effective mandibular length. Turner syndrome patients showed wide maxillary sinus, maxillary retrognathia and increased labio-inclination of lower central incisors to their apical base.

INTRODUCTION

Noonan and Turner syndromes are characterized by similar phenotypes, mainly short stature. Noonan syndrome was first described by Kobylinski, 1883[9], and later differentiated from Turner syndrome by Noonan and Ehmke in 1963[12]. It is an autosomal dominant disorder[17] and its incidence has been estimated to be between 1/1000 and 1/2500 live births[12]. It affects both sexes; some cases occur in sibs who are sometimes born to consanguineous parents[1]. Cytogenetic studies in Noonan syndrome revealed normal chromosomes[11]. It shares Turner stigmata with cardiac defects and a tendency towards mental retardation[18].

Turner syndrome was described in 1938. Its incidence is approximately 1/10000 females. Chromosomal findings are 45,XO or equivalents or mosaics such as 45,XO/46, XXp;46,XX/45,XO; 46XXi[18]. Cardinal features include short stature, streak gonads with sexual infantilism, somatic abnormalities, low birth weight, triangular face, prominent ears, webbed neck, low posterior hair line, multiple pigmented neavi, shield chest, lymphedema, hyperconvex nails, cubitus valgus, renal abnormalities, congenital heart disease, cognitive defects and autoimmune problems[2].

It is thus important to document and compare the oro-dental and craniofacial manifestations of both Noonan syndrome and Turner syndrome clinically and radiographically, to assess the important differences and similarities that could be of diagnostic value in both syndromes.

SUBJECTS AND METHODS

In the present study, three groups were included for comparison group (I) 10 normal control females of similar age to the second and third groups, group (II) 10 cases of Noonan syndrome and group (III) 10 cases of Turner syndrome.

All studied cases were subjected to pedigree analysis, clinical examination, cytogenetic findings, clinical photographs and radiographic examination (including both lateral and postero-anterior cephalograms and panoramic radiographs).

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