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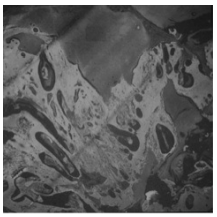
Case Report

Turner syndrome with dental structural abnormalities: histological and morphostructural features by confocal laser microscopy



A.P. Cazzolla[1], F. Riccitiello[2], V.A. La Carbonara[1], S. Franco[1], M.G. Lacaita[1]

[1]Clinic of Dentistry, University of Bari, Italy; [2]Department of Neuroscience, Reproductive Sciences and Dentistry, University of Naples "Federico II", Italy

Turner syndrome is a genetic disorder characterized by the total or partial absence of the X chromosome. There are different types of karyotypes although the most frequent is the 45X (57%). Mosaicisms such as 46 XX/45X, 45X/46XX/47XXX (29%), the presence of the isochromosome X or the chromosome X-ring (14%) may occur.

The main clinical features are: short stature, gonadal dysfunction, congenital heart disease, renal and skeletal anomalies, endocrine disorders and hypoplasia of the fingernails and toenails. Craniofacial features are due to retarded development of the cranialfacial complex with reduced growth of the skull base, hypertelorism, retrognathic profile, malocclusion Class II (60%), hypoplasia of the maxilla, bilateral cross-bite (9%) with or without cleft palate, anterior open bite (17%). The teeth are altered in shape and structure.

A patient AA 9 years old, suffering from Turner syndrome karyotype 45Xq; the Rx OPT has highlighted the presence of agenesis of 2.2 and a mesiodens in the region between the upper central incisors with the inclusion of 1.1. We proceeded to the extraction of the pre-orthodontic mesiodens and subsequent orthodontic treatment for the correction of Class II malocclusion.

The mesiodens, fixed in formalin, decalcified, and half was embedded in paraffin and stained with hematoxylin-eosin and half including resin-ground sections. Histological examination to the confocal laser scanning microscopy showed:

- Alternation of zones of hypo and over calcification;
- Inhomogeneous structure of dentin and abnormalities of the dentinal tubules;
- Reduced enamel thickness and irregular arrangement of the striae of Retzius;
- The pulp and root systems are characterized by the presence of foci of ectomesenchymal dentinogenetic multifocal differentiation that cause irregularities in the shape and structure of the root by an irregular, neoplastical-like growth (odontoma-like).

References

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