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Gorlin-Goltz syndrome: Role of the Orthodontist in clinical and radiological diagnosis

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Background and aim: Gorlin-Goltz syndrome is an autosomal dominant inherited disease. Typical manifestations involve the maxillary district, with the presence of keratocystic lesions, basal cell carcinomas and skeletal deformities. It is necessary to promptly recognize the pathology in order to improve the patient's quality of life, through a multidisciplinary approach that actively involves the orthodontist.

Materials and Methods: A systematic review of the literature was conducted through PubMed database, to identify diagnostic radiological criteria that could place Gorlin-Goltz syndrome in the differential diagnosis.

Results and conclusions: About 60% of patients present dysmorphisms such as macrocephaly, bulging forehead and facial rila. Skeletal anomalies such as fused or cuneiform vertebrae, hemivertebrae, kyphoscoliosis may be present; Presence of facial dysmorphisms such as cleft lip / palate, macrocephaly and ocular anomalies. The maxilla may appear hypoplastic and mandibular hyperplasia with variable prognathism may be present. Other less frequent skeletal anomalies are malocclusion and dental crowding, caused by the presence of keratocysts that can cause dislocation of the dental elements, non-eruption and root resorption. Typical radiographic findings in the jaws are multilocular radiolucent lesions, with well-defined sclerotic edges. Keratocysts can become symptomatic in the event of infection, nerve compression, tooth mobility or edema; They are more localized in the area of the mandibular ramus, while the mandibular body and the upper jaw are less frequently affected areas. Keratocysts are usually surrounded by a few satellite cysts. This characteristic favors a high recurrence of the lesion after surgical removal, therefore a follow-up with radiographic examination is indicated. Other typical lesions can be found images of the skull, where bilamellar calcification of the falx cerebri and alterations of the sella tucica are occasionally found (4). The orthodontist plays a fundamental role in the therapeutic and diagnostic plan of patients suffering from GGS, since through the periodic radiographic examination it is possible to identify early predictive lesions and set up a treatment plan for the resolution of skeletal and dental anomalies and a follow up for monitoring of relapses.

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