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Oral Pathology

Cherubism

Cherubism is a genetic disorder that only affects jaw bone, with features of bilateral swelling of the cheeks. It is an autosomal dominant disease and may affect multiple members of a family. Radiographic appearance of the lesion is describe as soap bubble appearance or multilocular radiolucency.

The disorder is caused by a point mutation in the SH3BP2 gene located on the chromosome 4p16.3, this leads to the malfunction of the Msx-1 gene, which is "responsible for mesenchymal interaction in craniofacial morphogenesis." In a simpler explanation, the disorder is caused by proliferation of fibrovascular tissue containing multi-neucleated Giant cells. The disorder can develop without a family history of the condition. The disease is often recognized in age two to five years of life, and is associated with bone reabsorption and fibrous expansion continuing into puberty. The fibrous tissue can expand the cortical bone which cause the swelling in the mandible. In severe cases the expanding masses in the maxilla bulges in to the orbital floor, this cause an upward tilting of the eyes.

Cherubism clinically can be describe as painless bilateral bony swelling of the posterior mandible and maxilla. The patient can present with broad cheeks, round face and even prognathism. The mandible is mostly affected especially areas such as the retromolar/molar area and the ascending rami. The lesion is unencapsulated and with no periosteum. The consistency

can range from semi hard to jelly like. The lesion is associated with dental anomalies, such as misaligned erupted teeth, missing mandibular third and second molar. The younger the lesion surface, the faster the lesion progress. The growth of the lesion can lead to deformation of facial bone and respiratory distress or even impaired vision or hearing. The lesion is twice in common in boys than in girls and is a childhood pathology.

Radiograpically the lesion has a soap bubble appearance, it is bilaterally symmetrical, well-defined multilocular radiolucencies in the mandible and sometimes in the maxilla. When a biopsy is conducted, there are giant cells throughout connective tissue. There is no need for active treatment of the pathology, because the patient is followed for stabilization and the lesion tends to go through spontaneous regression. Maxillary lesions then to regress faster than mandibular lesion. Sometimes it can treated with removal of impacted teeth or curettage of fiberous tissue. If surgery is required, it is best done after puberty, because early surgery can predispose for recurrence of the lesion. Cherubism can be mistaken with Giant cell tumors, such a Central Giant Cell Granuloma.The disease is relevant to me as a dental hygienist because, as a future dental hygienist , I might see a patient who comes for a cleaning presents with these clinical symptoms for Cherubism. It is good to know how Cherubism presents clinically by looking at the patient facial profile or intra orally; therefore I would to be able to identify the oral pathology, that is Cherubism so the patient can seek help.

Reference

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