

# Maxillofacial Radiology 198

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## CASES

Two paediatric male patients presented with painless bilateral facial swellings (Figures 1 and 2A&B). What are the pertinent radiological findings and your diagnostic hypothesis?



Figure 1: Panoramic radiograph of patient 1

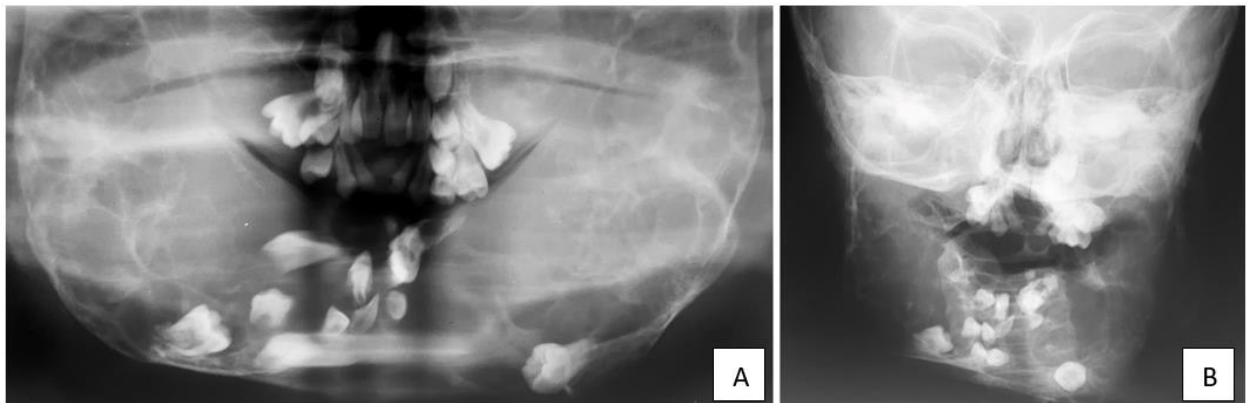


Figure 2: Panoramic radiograph (A) and Posterior anterior skull radiograph (B) of patient 2

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Liam Robinson: 50%  
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## INTERPRETATION

Both patients presented with bilateral multilocular radiolucent lesions that were highly expansile with varying degrees of tooth displacement, including multiple impactions. In case 1 the lesions were limited to the mandible, whereas case 2 presented with extensive involvement of both jawbones.

Cherubism is a rare bone disorder that may be inherited in an autosomal dominant pattern (68% of cases) with varying penetrance, or in rarer instances may occur sporadically. The aetiopathogenesis involves a mutation of the SH3 domain-binding protein 2 (*SH3BP2*) resulting in extensive giant cell lesions replacing bone.<sup>1</sup> *SH3BP2*-

dependant signal transduction seems to be involved in the regulation of osteoclastic and osteoblastic activities.<sup>2</sup> Clinically, patients present with symmetrical expansion of the jaws that characteristically undergo regression after puberty. Maxillary involvement may lead to displacement of the orbital floor giving an 'eyes turned to heaven' or 'plumped-cheek angel' appearance. Cherubism shows a slight male predominance, presenting at a mean age of 6 years. Radiologically, cherubism presents with highly characteristic features of bilateral well-defined multilocular radiolucent lesions with significant bony expansion. Tooth displacement (95%), tooth agenesis (62%), root resorption (40%) and cortical destruction (35%) are also commonly seen.<sup>1</sup> The second and third molars are frequently absent, as noted in case 2.<sup>2</sup> This could be explained by the correlation between the timing of development of cherubism and the associated missing molars. Others speculate that this presentation may be related to the pathogenesis, as a genetically determined alteration of tooth germ development.<sup>2</sup> Unilateral cases have been reported, however strict clinicopathological workup is required to rule out other differential diagnoses. Due to spontaneous regression after growth, longitudinal observation is the suggested management modality. Numerous drugs including bisphosphonates, calcitonin,

interferon, and imatinib, amongst others, have been used with varied outcomes. Currently, due to limited studies on these therapeutic approaches, conclusions on their effectiveness cannot be drawn. Surgical intervention is only indicated in the presence of functional or aesthetic impairments, as some cases treated via surgical intervention resulted in rapid regrowth.<sup>1</sup>

#### REFERENCES

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