New Oral Findings in Hamamy Syndrome

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Hamamy syndrome is a rare autosomal recessive disorder first described in 2007 in a Jordanian family (Hamamy et al, 2007). The syndrome is caused by missense mutations in the IRX5 gene on chromosome 16q12.2-q21 (Bonnard et al, 2012). Clinical findings in Hamamy syndrome include sever hypertelorism, prominent midface, prominent ears, sever myopia, learning disability, and multiple fractures due to bone fragility.

Oro-dental findings in Hamamy syndrome are not well documented in the literature, but reported features include loss of lamina dura, deep palate, enamel hypoplasia and dilacerated roots (Guler & Keskin, 2014; Hamamy et al, 2007). We report, for the first time, the occurrence of dentinogenesis imperfecta type III (Brandywine variant or shell teeth), tooth impaction, and multiple gingival polyps in Hamamy syndrome. A 14-year old girl known to have Hamamy syndrome presented to restore broken teeth. Extra-oral examination revealed characteristic features of the syndrome including severe hypertelorism (intercanthal distance= 6cm), broad nasal bridge, prominent ears with flat helix, and thin lips (Figure 1A). Upon intra-oral examination, severe coronal attrition was evident and the remaining tooth structure appeared grey-opalescent in colour (Figure 1B). Radiographic examination showed generalized large pulp chambers with a very thin layer of overlying dentine (i.e. shell teeth), and multiple impacted and displaced teeth (Figure 1C). The oral mucosa appeared normal, but three inflammatory gingival polyps (pyogenic granulomas) were evident on the lower and upper posterior gingivae (Figure 1B).

Recognition of dental abnormalities might help to differentiate Hamamy syndrome from other related disorders such as frontonasal dysplasia, craniofrontonasal syndrome, and Teebi hypertelorism syndrome. Further studies are needed to report the dental abnormalities in patients with Hamamy syndrome.

References

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Figure legends:

Figure 1A: Characteristic facial features in Hamamy syndrome

Figure 1B: Severe attrition and grey-opalescent teeth

Figure 1C: Enlarged pulp chambers and thin dentine (i.e. shell teeth)

Author contribution:

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