Journal of Dental Sciences 17 (2022) 1407–1408

Available online at www.sciencedirect.com

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journal homepage: www.e-jds.com

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Gorlin syndrome – Case report

KEYWORDS
Gorlin syndrome;
Nevoid basal cell carcinoma syndrome;
Odontogenic keratocyst;
Calcification of the falx cerebri;
Palmar or plantar pits

Gorlin syndrome is also called as nevoid basal cell carcinoma syndrome.[1–4] Although the patient with Gorlin syndrome may have many characteristic clinical features, in dentistry the Gorlin syndrome is always diagnosed initially by the presence multiple odontogenic keratocysts (OKCs). Here, we reported a case of Gorlin syndrome with multiple OKCs in the jawbones of a 21-year-old male patient.

This 21-year-old male patient had a past medical history of cleft lip and palate which had been treated by surgery with a relatively good clinical outcome. He came to our dental clinic for treatment of the right facial swelling with cellulitis for a week. Panoramic radiography revealed a well-defined multilocular radiolucency with corticated margin at the right posterior maxilla, extending from tooth 16 to the pterygoid plate and measuring 2.8 × 2.1 cm in size. The tooth 17 was included in the radiolucent lesion and tooth 18 and the right sinus floor was pushed upward (Fig. 1A). Moreover, another well-defined multilocular radiolucent lesion with corticated margin was discovered at the left posterior maxilla, extending from tooth 26 to the pterygoid plate and measuring 2.8 × 2.1 cm in size. The tooth 27 and the left sinus floor was pushed upward (Fig. 1A). The posterior-anterior (PA) skull film revealed calcification of the falx cerebri (Fig. 1B). In addition, the patient also had mild ocular hypertelorism, bifid rib, palmar/plantar pits, and short-ended metacarpals (data not shown). Under the clinical impression of Gorlin syndrome and the right facial cellulitis, he was admitted three days later. Furthermore, the enucleation of the bilateral maxillary cystic lesions and removal of the impacted teeth 17, 18, and 27 were performed under general anesthesia. Approximately 4.5 months later, the patient came to our clinic with the chief complaint of pain and swelling at the bilateral posterior mandibular gingival areas. Panoramic radiography revealed two radiolucent lesions at the distal regions of teeth 37 and 47 (data not shown). These two radiolucent lesions were also enucleated under general anesthesia. All the four cystic lesions showed the same characteristic histopathological features of OKCs. Microscopically, the cystic lesion was lined by a thin layer of parakeratotic epithelium with palisaded columnar basal cells (Fig. 1C, D and E). Budding of the basal epithelial cells to form small epithelial islands into the underlying connective tissue wall was discovered (Fig. 1F and G). Moreover, odontogenic epithelial rests were also observed in the fibrous cystic wall (Fig. 1H). Therefore, the characteristic microscopical findings confirmed the diagnosis of OKC for all four cystic lesions in the maxilla and in the mandible of our patient.

There are five major diagnostic criteria for the Gorlin syndrome, i.e., 1) five or more basal cell carcinomas or one before the age of 30 years, 2) OKC, 3) lamellar calcification of the falx cerebri, 4) two or more palmar or plantar pits, and 5) first degree relative with the Gorlin syndrome.[1–4] Because our patient had 3 of 5 major diagnostic criteria, there is no doubt to make the diagnosis of Gorlin syndrome.

https://doi.org/10.1016/j.jds.2022.05.004

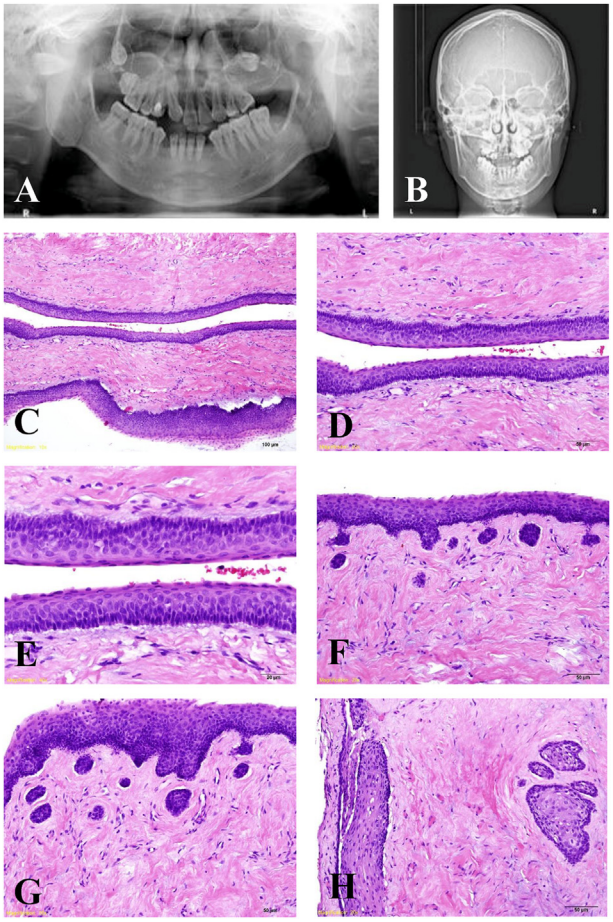

Figure 1 Radiographic and histopathological photographs of the patient with the Gorlin syndrome and odontogenic keratocysts. (A) Panoramic radiograph revealing two well-defined multilocular radiolucent lesions with corticated margin and impacted teeth 17, 18, and 27 at the bilateral posterior maxilla. (B) The posterior-anterior skull film showing calcification of the falx cerebri. (C, D and E) Medium and high-power microphotographs exhibiting a cystic lesion lined by a thin layer of parakeratotic epithelium with palisaded columnar basal cells. (F and G) High-power microphotographs showing budding of the basal epithelial cells to form small epithelial islands into the underlying connective tissue. (H) High-power microphotograph showing odontogenic epithelial rests in the fibrous cystic wall. (Hematoxylin and eosin stain; original magnification; C, 10 × ; D, 20 × ; E, 40 × ; and F, G and H, 20 ×).

Because the development of the basal cell carcinoma seems to be triggered by ultraviolet light exposure, patients should take appropriate precautions to avoid sunlight.[1] In addition, a close follow-up of the patient is absolutely needed.

Declaration of competing interest

The authors have no conflicts of interest relevant to this article.

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Received 6 May 2022
Available online 21 May 2022

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