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Case Report

Impacted tooth extraction from osteoma in a patient with Robinow syndrome diagnosed from intraoral lesions: A case report and literature review

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ABSTRACT

Robinow syndrome is very rare and is characterized by congenital anomalies of the head and neck. Symptoms occurring in the head and neck region include osteosclerosis of the skull, midface hypoplasia, cleft palate, and impacted teeth. There is no case report describing oral maxillofacial surgery in Robinow syndrome. We report the case of a 19-year-old female in whom a panoramic radiograph revealed a radiopaque lesion in the right maxillary molar region along with multiple impacted teeth, hypodontia, and retained deciduous teeth in her jaws. Impacted tooth was also present in the radiopaque lesion on the right maxilla. Computed tomography revealed a high-density area on the right maxilla, representing a lesion containing an impacted tooth. The general findings and the presence of impacted teeth indicated symptomatic disease, and biopsy examination confirmed an osteoma. After additional examinations, Robinow syndrome was diagnosed. Tooth extraction was performed under general anesthesia. About 8 months after operation, the wound showed epithelialization and osteoma-like tissue formation was confirmed in the extraction socket. We considered repeated observations to be necessary in this case because of the possibility of recurrence. It is necessary to be aware of the possible diagnosis of Robinow syndrome on the basis of intraoral symptoms after school age.

1. Introduction

In 1969, Robinow described a syndrome of characteristic facies (fetal face), mesomelic limb shortening, and external genital hypoplasia [1,2], now known as Robinow syndrome (RS). Symptoms occurring in the head and neck region include osteosclerosis of the skull, midface hypoplasia, cleft palate, and impacted teeth.

There is no case report describing oral maxillofacial surgery in RS. We report a case of impacted tooth extraction from an osteoma of a patient with RS diagnosed on the basis of intraoral lesions.

2. Case report

A panoramic radiograph at the orthodontics clinic revealed a radiopaque lesion in the right maxillary molar region in a 19-year-old female. She was admitted to our department for detailed examination and an orthodontic surgery consultation by the primary care orthodontist in October 2012.

At birth, she was found to have hypertelorism and nasal septum cartilage hypoplasia. She was evaluated for suspected hydrocephalus and Down syndrome, but there were no abnormal findings. Her family history was unremarkable, and her parents' marriage was not consanguineous. At the age of 6 years, she underwent umbilicoplasty and coccyx osteoplasty; and at the age of 16 years, she underwent nasal septoplasty.

At the time of first visit to our department, her body mass index was 28.0 (height 150 cm, weight 63 kg). She had brachydactyly (Fig. 1a). Her facial findings comprised low-set ears, hypertelorism, hyperplasia of the skull, and concavity of the midface (Fig. 1a and b). An oral examination revealed bifid uvula and ankyloglossia (Fig. 2a and b). The right maxillary molar region showed

☆ Asian AOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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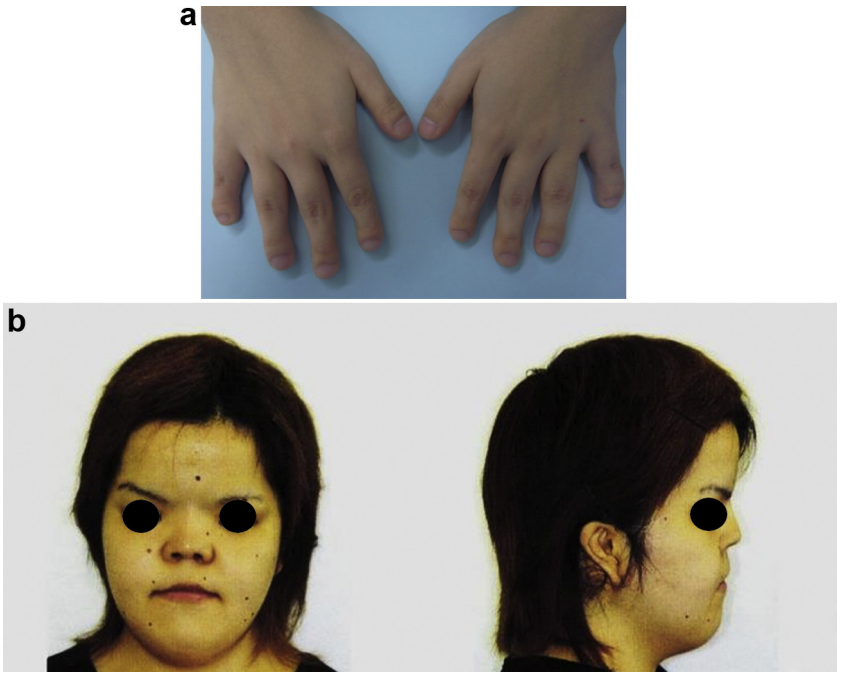

Fig. 1. (a) The patient had brachydactyly. (b) Facial findings comprised low-set ears, hypertelorism, hyperplasia of the skull, and concavity of the midface.

diffuse bone bulging. Most of her teeth showed enamel hypoplasia (Fig. 2c–e).

A panoramic radiograph revealed multiple impacted teeth, hypodontia, and retained deciduous teeth in her jaws. The radiopaque lesion on the right maxilla contained impacted tooth (Fig. 3a). Computed tomography (CT) revealed a high-density area in the right maxilla; the lesion contained an impacted tooth that was close to the right nasal cavity (Fig. 3b–d). Lateral cephalometry showed midface hypoplasia and osteosclerosis of the skull (Fig. 4a). Bone scintigraphy with ^{99m}Tc-MDP showed an increase in radioisotope (RI) uptake in the skull. RI uptake in the maxilla, mandible, and osteoma was within the normal range (Fig. 4b).

In October 2012, a biopsy of part of the right maxillary lesion revealed a pathologic diagnosis of osteoma. We had suspected Gardner syndrome because of the impacted teeth and osteoma. Further systemic examinations excluded Gardner syndrome. We suspected another symptomatic disease from the general and oral findings. We consulted other pediatric staff at our hospital, and RS was diagnosed by clinical geneticists.

We planned Le-Fort I osteotomy, but we expected that the impacted tooth could be an obstacle in this surgery. After discussion

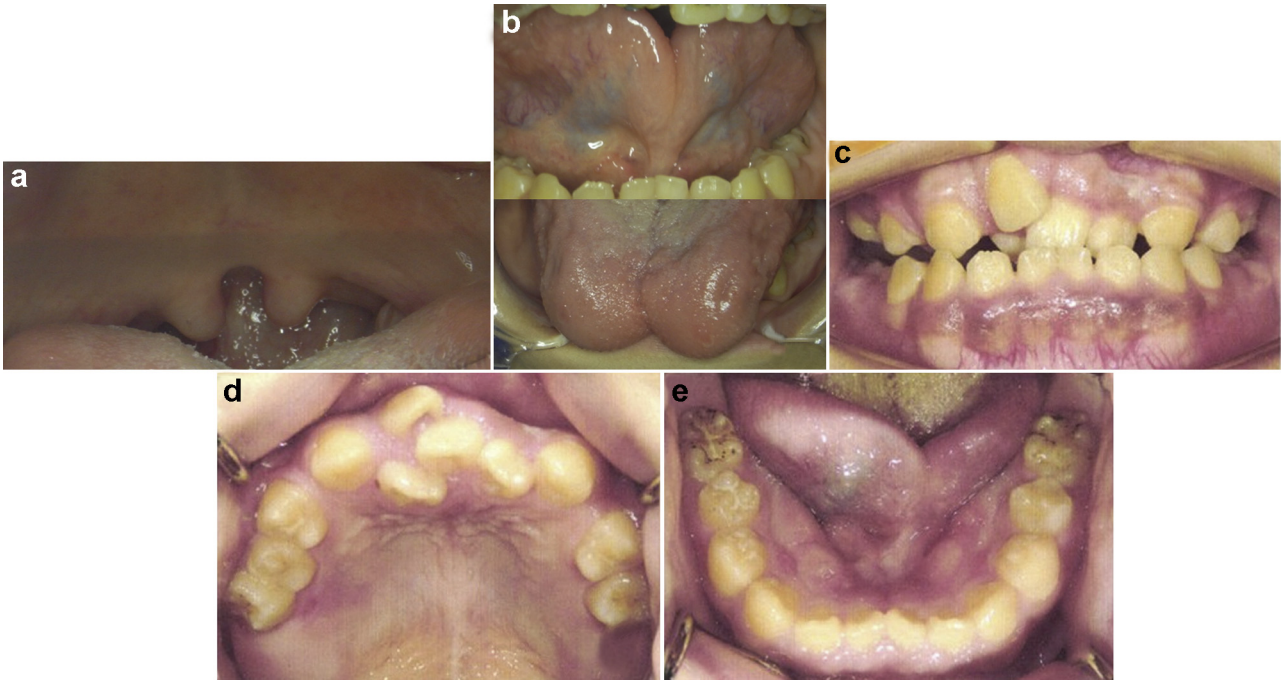

Fig. 2. (a) Bifid uvula. (b) Ankyloglossia. (c–e) Most of the teeth showed enamel hypoplasia.