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Case report

Congenital parotid ectopia in accessory maxilla and facial cleft anomalies: Three cases report

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ABSTRACT

To further document the clinical features of accessory maxilla with three additional cases report. Clinical and radiological features of three cases of accessory maxilla were presented. Related literature was summarized for comparison. Ectopic parotid gland, facial cleft and accessory maxilla are three concomitant malformations in this condition. The tooth-bearing accessory maxillary duplication derives from the abnormal growth of the zygoma or zygomatic arch. Facial cleft, parotid ectopia and tooth-bearing accessory maxilla may constitute a rare congenital syndrome.

1. Introduction

The terms of "maxillary duplication" and "accessory maxilla" have been used to describe an extremely rare clinical entity characterized by redundant tooth-bearing bony segment posterior to the maxillary tuberosity mimicking an accessory maxilla [1–3]. Up to now less than 20 cases have been documented in the English literature [1–14].

Cameron described the following clinical characteristics of this congenital anomaly: an accessory tooth-bearing maxilla arising from the inferior border of zygoma; supernumerary teeth of normal morphology in the accessory maxilla; full complement of maxillary dentition; facial cleft [8].

Ectopia of the parotid gland has not been reported in this malformation. CT imaging allows for radiological diagnosis of bone and parotid malformations [15]. We present three additional cases to illustrate the parotid malformation in this circumstance.

2. Case report

This study was approved by the Institutional Review Board of School and Hospital of Stomatology Peking University (PKUSSIRB-2012084).

2.1. Case 1

A 13-year-old boy with facial deformity was referred to our hospital. The patient was discovered with bilateral lateral facial clefts and cleft palate at birth. Surgical repair of the facial clefts and palatoplasty were performed at 5 years old. Familial history was unremarkable.

Physical examination showed the bilateral facial scars due to the surgical repair of the lateral facial clefts (Tessier 7). Cheek prominence and shallow grooves extending from the facial scars to the bilateral zygomata were present. Hypoplasia and retrognathia of the mandible was remarkable. Cleft soft palate was found and hypernasality was prominent. Posterior to the maxillary tuberosity on each side was an abnormal segment of bone (Fig. 1). Mucosal sulcus between the accessory maxilla and the maxillary tuberosity on each side could be seen. Bilateral buccal mucosa scars and folds existed and the openings of the Stensen's ducts could not be identified. Panoramic radiography revealed two erupted and three impacted supernumerary molar-like teeth in the right accessory bone segment. One erupted tooth and another impacted molar-like supernumerary tooth were found in the left accessory bone segment (Fig. 2). Another conical supernumerary tooth was observed in the left tuberosity (Figs. 1 and 2).

CT showed that the accessory teeth-bearing bony segments extended from the inferior borders of bilateral zygomata to the maxillary tuberosities (Fig. 3A). A narrow bony gap was observed between the normal maxilla and the accessory maxilla on the left

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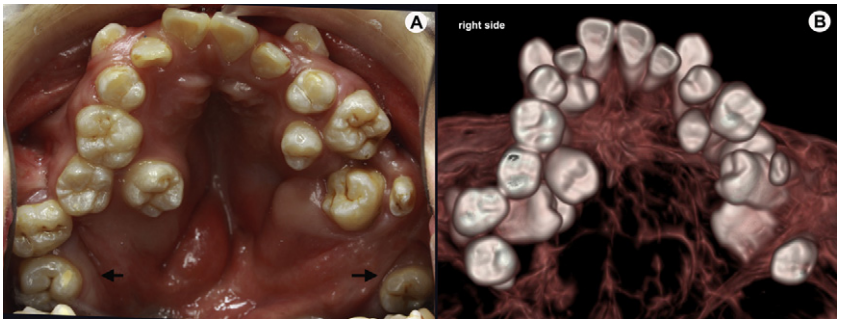

Fig. 1. Intra-oral clinical (A) and three-dimensional CT (B) views of the maxillary dentition. Posterior to the maxillary tuberosities on each side is the accessory maxilla. Two (right side) and one (left side) erupted, three (right side) and one (left side) impacted supernumerary teeth with molar shape could be observed in the accessory bone. Another erupted conical supernumerary tooth is observed on the left tuberosity. Note the narrow mucosal sulcus between the maxillary tuberosity and accessory maxilla on each side (black arrows).

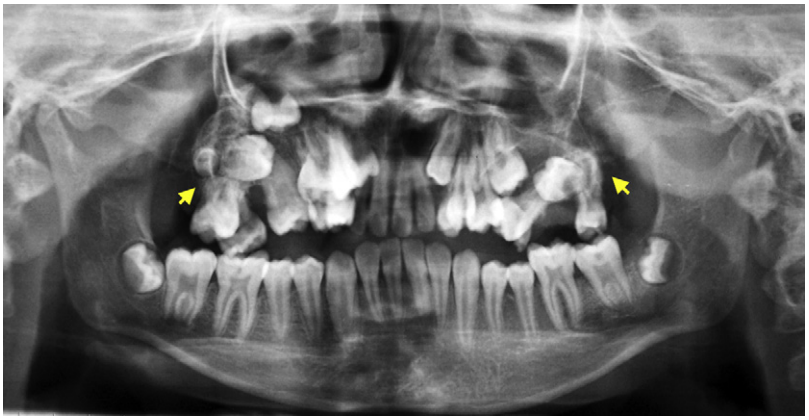

Fig. 2. Panoramic radiograph shows bilateral accessory maxillae (yellow arrows) with multiple teeth posterior to the maxillary tuberosity and the full complement of the permanent maxillary dentition. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

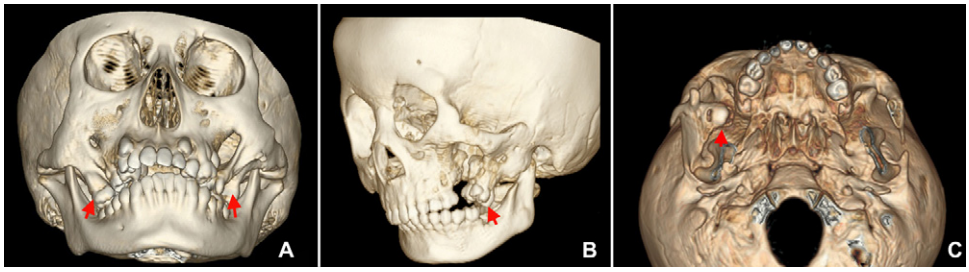

Fig. 3. Three-dimensional CT images. (A) Bilateral accessory maxillary bone segments extend from the inferior borders of bilateral zygoma to the maxillary tuberosity; (B) the accessory maxilla runs from the inferior border of the zygomatic process of the temporal bone to the maxillary tuberosity. Several supernumerary teeth are present in the accessory maxilla. Note the full complement of the primary maxillary and mandibular dentition; (C) malformations of bilateral zygomatic arches. Note the supernumerary tooth on the right zygomatic arch (red arrow) and full complement of the primary maxillary dentition. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

side. The accessory maxilla attached to the maxillary tuberosity on the right side.

Soft-tissue CT images showed that the left parotid space was composed of adipose tissue except for the vessel structures. Interestingly, a 1.4 cm × 1.5 cm × 2.5 cm soft tissue nodule with gland attenuation was found below the zygoma, suggesting the existence of an ectopic parotid gland. The right parotid gland was normal (Fig. 4A).

The accessory maxillae were not excised. Distraction osteogenesis was carried out to increase the mandible length. Orthodontic treatment was performed and functional occlusion was acquired.

2.2. Case 2

A 4-year-old girl was referred to our hospital because of facial deformity. She had undergone cheiloplasty because of congenital left macrostomia (Tessier 7). Physical examination showed left hemifacial microsomia and cheek prominence. The left outer ear was morphological normal but a few centimeters lower compared with the right ear. Oral examination revealed that an accessory bone segment with several supernumerary molars located posterior to the normal primary dentition on the left side. Mucosa sulcus was seen between the accessory bone and the maxillary tuberosity. The opening of the left Stensen's duct could not be identified and no obvious excretion could be elicited.

Three-dimensional CT showed that the accessory maxilla extended from the zygomatic process of the temporal bone toward the maxillary tuberosity (Fig. 3B). Four supernumerary molar-like teeth were observed in the accessory maxilla. Hypoplasia of the left mandibular coronoid process was also noted. The left parotid space was composed mainly of adipose tissue (Fig. 4B). A 2.0 cm × 1.5 cm × 1.0 cm soft tissue nodule of

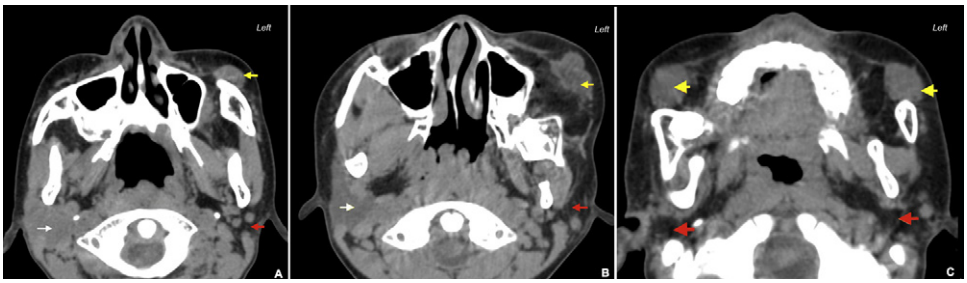

Fig. 4. CT manifestations of ectopic parotid gland. The parotid glands are absent in the left retromandibular parotid spaces in case 1(A, red arrow) and case 2 (B, red arrow). Bilateral parotid glands in the parotid space are absent in case 3 (C, red arrows). The parotid spaces without gland tissue are composed of fat tissue except for the areas of the retromandibular vein and internal maxillary artery. The ectopic glands locate slightly inferior to the ipsilateral zygomata (A–C, yellow arrows). Note the normal parotid glands in the right parotid spaces in cases 1 and 2 (A and B, white arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

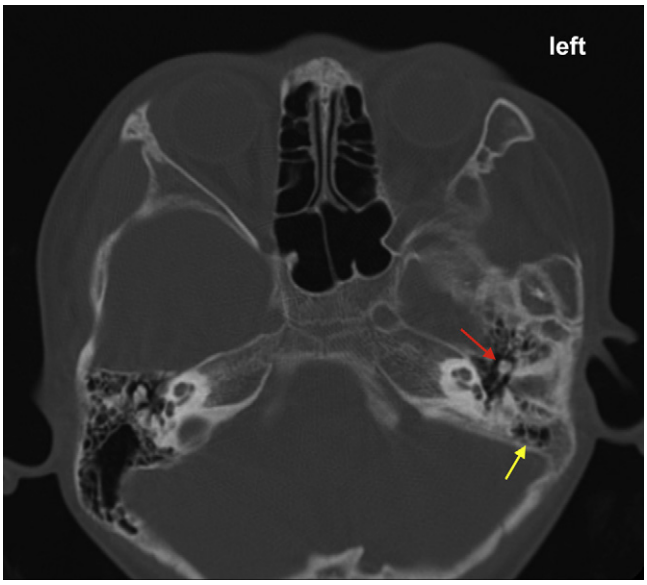

Fig. 5. Axial CT of case 2 shows the hypoplasia of the temporal bone (left side). Note the ossicular deformity (red arrow), poor pneumatization of the tympanic cavity and the mastoid air cells (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

gland attenuation was found below the zygoma, suggesting the existence of an ectopic parotid gland. The external acoustic canal was inferiorly oriented and stenosed (Fig. 5). Abnormal rotation of the malleus and incus and hypoplasia of the incus was observed.

The accessory maxilla was excised under general anesthesia and the recovery was uneventful.

2.3. Case 3

A 5-month-old boy with congenital macrostomia was referred to our hospital. Physical examination showed the lateral facial cleft approximately 1 cm in length on the right side (Tessier 7). No other finding was remarkable. The patient underwent cheiloplasty at 5 months old. The patient was recalled for examination at 2 years old. Bilateral zygomatic arch slightly enlarged and abnormal bony prominence could be palpated posterior to bilateral maxillary tuberosity. Full complement of primary dentition could be observed. Three-dimensional CT images showed that bilateral zygomatic arches were malformed due to abnormal downward overgrowth (Fig. 3C). A tooth bud was present in the abnormal bone segment on the right side. Bilateral parotid glands in the main parotid spaces were replaced by fat tissue. Soft tissue masses, approximately 1.5 cm × 2.0 cm × 2.0 cm in size, with gland attenuation indicating ectopic parotid glands were found below bilateral zygomata (Fig. 4C). The patient was kept in close follow-up.

3. Discussion

In the literature review, we searched for the congenital deformity featured by the presence of an accessory tooth-bearing maxilla posterior to the maxillary tuberosity. Complete duplications of the palate, mandible or oral cavity indicating more severe forms of developmental disturbances were excluded [13,16–18]. As a result, 14 cases with a very similar set of clinical manifestations were retrieved (Table 1).

According to the best of our knowledge, the parotid malformations have not been documented in this entity [6,8]. Present case series found out that malformation of the parotid gland could frequently occur with maxillary duplication. In the case described

Table 1
Clinical features of tooth-bearing accessory maxilla in the literature.

Author	Sex	Age	Sides	Accessory maxilla	Supernumerary teeth	Maxillary dentition	Facial cleft (Tessier no.)	Other findings
Borzabadi-Farahani [3]	F	7 y	L&R	Extended to maxillary tuberosity	4 molar-like teeth on both sides	Full crowded	None	Small mandible
Morita [1]	F	2 m to 22 y	L	From the zygomatic arch	Multiple molar-like teeth	Right alveolar cleft	None	Cleft soft palate
Sjamsudin [2]	F	3 y	R	From right zygoma	Incomplete deciduous dentition	Normal	Macrostomia Tessier 7	Cleft soft palate; microglossia
Tharanon [4]	M	12 y	L&R	Extended to maxillary tuberosity	Molars and molar buds	Full crowded	Bilateral Tessier 7	Macrostomia; retrognathia; cleft uvula
Jian [5]	F	17 y	R	Posterior to the premolars	2 premolars and 1 molar	Full crowded	Tessier 5	Defect of the right zygoma and maxilla
DeGurse [6]	M	6 y	L	Incorporated to the zygomatic arch	Multiple teeth, regular shape	Normal	Facial dimple lateral to the commissure	Mass in left pterygomaxillary fissure

Table 1 (*Continued*)

Author	Sex	Age	Sides	Accessory maxilla	Supernumerary teeth	Maxillary dentition	Facial cleft (Tessier no.)	Other findings
Miyajima [7]	F	4 y	L	Posterior to the maxillary tuberosity	3 molars	Normal	Tessier 5	Nasal deformity; cleft palate
Cameron [8]	M	6 y	L&R	From bilateral zygoma	Premolars and molars on both sides	Normal	Tessier 5	Cleft lip and alveolar crest
Cheung [9]	M	7 y	L&R	Fused to the skull base	7 and 9 teeth of regular shape on both sides	Normal	Tessier 7	Mandible hypoplasia, left ear deformity
Ryu [10]	M	4 y	L	Attaching to the skull base	4 deciduous molars	Normal	None	None
Ball [11]	F	4 y	L&R	Lateral to bilateral maxillae	Deciduous molars	Normal	Bilateral facial creases Tessier 7	Klippel–Feil syndrome; deafness
Stoneman [12]	F	11 y	L	Superior to the maxillary tuberosity	4 premolar-like teeth	Normal	Left congenital facial fistula	None
Smylski [13]	F	2 y	L&R	Extending to the skull base	3 teeth on the left side	Normal	Not described	Micrognathia
Rushton [14]	M	16 y	R	Attached to the sphenoid and deformed zygoma	4 molars or premolars on the right side,	Missing of a right molar	Tessier 7	Cleft uvulae; right lower-set ear
Present study	M	13 y	L&R	From bilateral zygomatic arches	5 teeth (right side) 2 teeth (left side)	Full crowded	Tessier 7	Micrognathia; left parotid ectopia; cleft palate
	F	4 y	L	From left zygomatic arch	5 teeth with molar shape	Normal	Tessier 7	Hypoplasia of mandible and temporal bone; left parotid ectopia
	M	2 m to 2 y	L&R	From bilateral zygomatic arches	Tooth buds on the right side	Normal	Tessier 7	Bilateral parotid ectopia

M: male; F: female; y: year; m: month; L: left; R: right; &: and.

by Morita [1], we could observe an abnormal round soft-tissue mass around the zygoma on the serial axial CT images, which showed similar appearance of ectopic parotid gland. However, the author did not give a soft-tissue view and detailed description.

As hypoplasia of the mandible, temporal bone, parotid gland, zygoma and facial cleft compose a regular set of manifestations, we suggest that it is a rare variant form of the first and second branchial syndrome.

CT is effective in identification of the developmental malformations of the parotid gland [15]. The missing of the gland tissue in the retromandibular parotid space could be replaced by fat tissue and the ectopic gland tissue could be found around the zygoma [15]. Ectopic parotid gland could occur due to the incomplete development of the gland. The ectopia of the parotid gland caused no symptom in present cases and no xerostomia was noticed.

Our CT findings also consolidated that the accessory maxilla extended from the inferior border of the zygoma or zygomatic arch [4,8]. Of utmost interest were the supernumerary teeth in the accessory maxilla, which frequently presented with the shape like molars or premolars. The teeth could establish occlusion with mandibular molars. The primary or permanent maxillary dentitions were normal or crowded in most cases. Mandibular hypoplasia, micrognathia and bony Class II malocclusion were common in this syndrome.

The amniotic band syndrome could possibly give rise to the craniofacial deformities including facial cleft, cleft lip and palate and micrognathia [7,19]. Fibrous bands due to premature ruptured amniotic sac could encircle and trap some part of the fetus, which will constrict and disturb the fetus growth. If an amniotic band is interposed between adjacent facial processes, it prevents fusion of those facial processes in early gestational age. This theory could well explain the concurrence of cleft palate or facial cleft defects [19] and incomplete formation of the parotid gland.

Neurocristopathy also manifests as orofacial clefts with maxillary duplication [5]. Neural crest cells are a transient population of multipotent precursor cells which give rise to diverse organs through embryogenesis. These cells become migratory and give rise to various tissues throughout the embryo. Abnormal neural crest cells migration could potentially results in abnormal growth of craniofacial cartilage and bone [4,5,8]. In this theory, the multipotent ectodermal epithelial bands in the primitive stomodeum responsible for tooth formation could give rise to abnormal migrations into the zygomatic anlage, which further lead to ectopic odontogenesis and osteogenesis.

Differential diagnoses include posterior alveolar cleft and teratoma. The presence of a sulcus in the posterior alveolar ridge could be easily misdiagnosed as posterior alveolar cleft clinically [7]. The presence of normal maxillary dentition could help differentiate the accessory maxilla from the alveolar cleft. CT could illustrate that the accessory maxilla extended from the zygomatic arch. Distinction should also be made between accessory maxilla and a teratoma containing bone and tooth structures. It is difficult if such a teratoma occurs immediately adjacent to the normal dentition, which may resemble an accessory maxilla. Supernumerary teeth in accessory maxilla were usually of regular molar and premolar shape. Eruption of these teeth and functional occlusion with mandibular teeth could be observed in accessory maxilla.

In conclusion, ectopia of the parotid gland is another anomaly in this rare congenital syndrome characterized by accessory maxilla and facial cleft.

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Contribution

All authors have made substantive contribution to this study and manuscript and all have reviewed the final paper prior to its submission.

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