Partial duplication of the mandible, parotid aplasia and facial cleft: a rare developmental disorder

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Duplication deformity of the mandible is exceedingly rare. Its occurrence with congenital facial cleft and parotid gland aplasia has been rarely reported as 1 entity. We report such a case with detailed computed tomography (CT) description and provide a review of the literature on mandible duplication. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;116: e202-e209)

Oral and maxillofacial duplication deformities vary from the fairly common (supernumerary teeth and branching of the mandibular canal) to the exceedingly rare (partial or complete duplication of the jaws).1 Duplication deformities of the mandible vary from symmetric doubled mandibular arches2 to partial duplication of individual structure of the mandible. Accessory condyle, coronoid process, mandibular body and canal may be present in the duplicated mandible.1,3,11 McLaughlin described the first case of reduplication of the mouth, tongue and mandible in 1948.4 Up to now, more than 10 cases of mandibular duplication have been documented in the English literature.2-14 Duplications of the maxilla have also been documented in several articles.15,16 This condition is believed to be a developmental malformation rather than teratoma. The pathogenesis is not clearly known.

Present report describes a rare case of partial duplication of the mandible. This case is characterized by the presence of an accessory tooth-bearing alveolar bone segment with duplicated ramus, mandibular foramen and canals. A transverse facial cleft from left commissure to cheek and aplasia of the left parotid gland are also present. The literature on mandible duplication has been reviewed for discussion.

CASE REPORT
A 15-year-old Chinese girl complaining of facial deformity was referred to our hospital. The patient was discovered with left facial cleft at birth and underwent surgical repair of the cleft when she was 2 years old. A photograph at the first surgery showed macrostomia, accessory mandible and an intraoral soft tissue band of the patient (Figure 1). The band was formed by the partially duplicated lower lip and separated the accessory mandible from the true mandibular teeth and tongue. The patient denied hearing loss or difficulty. Familial history and blood tests revealed no unusual finding.

Physical examination revealed a left facial scar due to the surgical repair of the transverse facial cleft (Figure 2). A non-tender bony prominence of the left mandibular body was palpated beneath the skin. Two skin dimples were discovered in the left preauricular region (Figure 2B). Intraoral examination showed that a soft tissue band extended from the left lower lip to the pterygomandibular raphe (Figure 3). The band was approximately 1 cm in width, non-tender and soft on palpation. Inferior to the soft tissue band was the prominently displaced permanent mandibular dental arch. Lateral and superior to the soft tissue band was an accessory bone segment with 2 erupted molar-like supernumerary teeth and contact overlying mucosa (Figure 3).

A panoramic radiograph (Figure 4) showed that the left mandibular dental arch was severely crowded and deformed due to the presence of the accessory teeth-bearing alveolar bone segment. Duplication of the coronoid process, ramus, and sigmoid notch were observed.

Three-dimensional volume rendering of a spiral CT study showed that the permanent mandibular left premolars and molars were displaced lingually and extended to the inner lesser ramus (Figures 5 and 6). The accessory alveolar bone segment was lateral to the permanent mandibular arch and extended to the true ramus. One impacted canine-like tooth and 5 molar-like teeth (2 erupted and 3 impacted) were observed in the accessory bone. Further observation of the mandibular canals was made by cone beam CT (Figures 7 and 8). The true mandibular canal entered the inner ramus (lesser one) through a regular mandibular foramen and exited the mandible via the mental foramen. Two redundant foramina were observed on the lingual side of the outer ramus (greater one) and opened into 2 redundant mandibular canals, which extended beneath the roots of the supernumerary teeth.

The soft tissue CT image (Figure 9) showed that the left parotid gland was absent in the parotid space. An ectopic parotid was found below the zygoma and superficial to the masseter. The right parotid gland was visually normal on CT.
Surgical excision of the accessory alveolar bone was performed under general anesthesia. After elevation of the mucoperiosteum flap, the accessory alveolar bone segment was chiseled from the outer aspect of the left body of the true mandible. The new mandible surface was shaped. The accessory coronoid process and ramus were not resected. The excised bone segment was composed of mature cortex and trabecular bone visually and pathologically. Six supernumerary teeth were extracted (Figure 10A). Radiographs of the 3 molar-like teeth showed relatively normal morphology of the pulp chambers (Figure 10B). One molar-like tooth presented with narrowed pulp chamber. The other 1 molar-like tooth fused with a cone-shaped tooth.

**DISCUSSION**

Craniofacial duplication is a rare form of conjoined twinning and presents in a wide spectrum, from diencephalus, diprosopus to partial facial duplication. Wu et al. reviewed the literature and reported that approximately 100 cases of complete or incomplete craniofacial duplication were identified.

Four types of mandibular duplications have been identified in the literature (Table I). Type I is characterized by symmetrically duplicated mandibular arches with deciduous teeth or tooth buds. Duplicate tongue, lip and cleft palate are present in this condition that may take the appearance of partially duplicated oral cavity. Type II is characterized by the duplication of the unilateral mandibular body and ramus. The duplicated mandible may extend from the symphysis to the temporomandibular joint as a separate hemi-mandible. Type III is the alveolar type, characterized by the presence of a localized accessory alveolar bone with supernumerary teeth attached to the normal mandible. The mouth may be partially duplicated to present as macrostomia or completely duplicated to present as a separate mouth in the type II and III deformities. The supernumerary teeth in the duplicated mandible are frequently of regular morphology. Type IV is characterized by the bilaterally...
duplicated ramus and its remarkable association with Klippel–Feil syndrome.\textsuperscript{10,12}

Facial cleft deformity is closely related to the duplicated mandible and maxilla.\textsuperscript{15-23} The transverse facial cleft in present case is considered to occur secondary to the duplication of the mandible and lower lip. The soft tissue band separating the normal dentition and the accessory mandible in present case is considered to be reminiscent of the duplicated lower lip, which is very similar to those described by Maisels and Suhaili.\textsuperscript{8,13}

Duplication of the ramus may be complete or partial in duplicated mandibles.\textsuperscript{3} In present case the duplication of the ramus is partial. The anterior of the ramus divides into 2 separated plates and gives rise to 2 coronoid processes and sigmoid notches. Redundant mandibular foramina and canals can be identified. Bilateral ramus duplication is related with the Klippel–Feil syndrome,\textsuperscript{10,12} which is characterized by congenital fusion of 2 or more cervical vertebrae and subsequent shortening of neck length and movement limitation.

This case adds to our knowledge in that parotid aplasia may be involved in the mandible duplication and facial cleft. Davies also reported an accessory salivary duct running from the duplicated mouth toward the anterior border of the masseter during surgical dissection.\textsuperscript{5} CT or MRI (magnetic resonance imaging) identification of the aplasia of the parotid gland is effective. The retromandibular parotid space is composed of fat connective tissue without the gland parenchyma in parotid aplasia.\textsuperscript{24} Ectopic underdeveloped parotid gland can be observed superficial to the masseter.\textsuperscript{24,25} Parotid aplasia in present case is quite similar to that described by Higley et al.\textsuperscript{24} Aplasia of 1 individual major salivary gland does not lead to significant clinical symptoms and does not necessitate any surgical intervene.

Duplication of the oral and maxillofacial structures has been interpreted due to a variety of different pathogenesis: (1) forking of the notochord, (2) duplication of the prosencephalon, (3) duplication of the olfactory placodes, (4) duplication of the maxillary or mandibular growth centers around the margins of the stomatodeal plate.\textsuperscript{26}
Fig. 6. Three-dimensional CT images of the mandible show that an outer greater coronoid process (white arrow, A) and an inner lesser coronoid process (red arrow, A) are present. Note the mandibular foramen (blue arrow, A) opening into the internal side of the ramus. The permanent mandibular left premolars (red arrows in B) and molars (black arrows in B) are displaced lingually and extend to the inner lesser coronoid process. The supernumerary teeth (white arrows in B) are of molar’s morphology and run to the outer greater coronoid process.

Fig. 7. Axial cone beam CT images (A-B) showing the structure of the duplicated ramus. The ramus is partially duplicated to form an outer (white arrow, A) and inner (black arrow, A) coronoid processes. The 2 processes fused together at the posterior border of the ramus (white arrow head, A). The inherent mandibular foramen (black arrow, B) is observed on the internal side of the inner ramus and opens into the inherent mandibular canal. On the internal side of the outer coronoid process, 2 redundant foramina (white arrows, B) opening into 2 individual bony canals (white arrows, C) toward the accessory alveolar bone are observed. Oblique reformatted image (C) shows the redundant foramina and canals in 1 image (white arrows).

Fig. 8. Oblique reformatted cone beam CT images showing the inherent mandibular canal (black arrow, A) and redundant mandibular canals (white arrows, A-B).
McLaughlin suggested that it was the result of reduplication of certain elements derived from the first branchial arch.\(^4\) Split notochord theory could best illustrate the embryogenesis of various degree of oral and facial duplications.\(^2\,^8\) Davies also regarded it as a developmental anomaly arising from separated totipotent cells.\(^5\) Similar neurocristopathy theory may also well explain the occurrence of orofacial clefts with maxillary duplication.\(^15\,^20\) This theory can explain how duplicate oral, maxilla or mandible occur, but cannot explain how parotid gland developmental dysplasia occurs in the present case.

Amniotic band syndrome has been suggested to be correlated with facial cleft deformity.\(^21\,^27\) The amniotic band syndrome is due to a premature rupture of the amniotic sac. Fibrous bands due to ruptured amnion can encircle and trap some part of the fetus, hence cause congenital abnormalities. If an amniotic band is interposed between adjacent facial processes, it prevents fusion of those facial processes in early gestational age and gives rise to facial clefts. It could also constrict and disturb the formation of the parotid gland. Mechanical restrictive force could reasonably distort the anlage of the mandible or the dental lamina to branch abnormally or migrate into redundant multicomponent cells, thus gives rise to various patterns of mandibular duplication.

Distinction should be made between accessory jaw and a teratoma containing osseous or tooth-like structures.\(^4\) A teratoma is an encapsulated tumor with various tissue or organ components deriving from 3 germ layers.

A teratoma may contain fat, hair, teeth and bone in disorganized arrangement. In contrast with teratoma, the accessory structures in duplicated mandibles are regularly organized. Supernumerary teeth in jaw duplication are usually of regular tooth shape. Eruption of these teeth and functional occlusion in duplicated jaws sometimes can be observed. Structural duplication of the mandibular structures including coronoid process and mandibular canals observed in duplicated mandible also helps differentiation.

In conclusion, the mandibular duplication can be partial and present with accessory tooth-bearing alveolar bone, doubled coronoid process and ramus. Facial...
### Table I. Summary of clinical characteristics of duplicated mandibles

<table>
<thead>
<tr>
<th>Author</th>
<th>Sex: age</th>
<th>Location</th>
<th>Type</th>
<th>Features</th>
<th>Supernumerary teeth</th>
<th>Other oral facial deformity</th>
<th>Other body site deformity (potential syndromes)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wittkampf and van Limborgh</td>
<td>F: 3 y</td>
<td>Symmetric duplication</td>
<td>I</td>
<td>W-shaped double mandible with osseous connection in the midline</td>
<td>Multiple deciduous teeth in the mandibles</td>
<td>Hypertelorism, macrostomia, duplicate lower lip, cleft palate, duplicate anterior tongue</td>
<td>Intraoral hamartoma, duplication of sella turcica and odontoid processes, fused vertebrae C2 and C3; (split notochord syndrome)</td>
</tr>
<tr>
<td>Wu et al.</td>
<td>M: newborn</td>
<td>Symmetric duplication</td>
<td>I</td>
<td>Two partially formed, separate mandibular arches</td>
<td>Teeth buds present in 2 mandibular arches</td>
<td>Duplicate tongue and upper labial frenulum; cleft palate</td>
<td>Exotropia; microphthalmus; generalized hypotonia; low-set ears; orbital hypertelorism; abnormal tectum; duplicate vertebral bodies from C2 to C5, C6 to T3</td>
</tr>
<tr>
<td>Mclaughlin</td>
<td>F: 3 m</td>
<td>Right mandibular body</td>
<td>II</td>
<td>Duplicated horizontal rami fused with the normal mandible to the left of the symphysis</td>
<td>Accessory mandible contained 8 teeth buds</td>
<td>Duplication of mouth and tongue</td>
<td>NS</td>
</tr>
<tr>
<td>Davies et al.</td>
<td>NS: 2 y</td>
<td>Right mandible</td>
<td>II</td>
<td>Accessory mandibular body fused with the enlarged ramus extending toward the temporomandibular joint</td>
<td>Eight supernumerary deciduous and permanent teeth with normal morphology</td>
<td>Duplicate mouth, accessory salivary gland duct</td>
<td>No</td>
</tr>
<tr>
<td>Shaikh et al.</td>
<td>F: 30 y</td>
<td>Left mandibular body and ramus</td>
<td>II</td>
<td>Duplication of the hemimandible, condyle, coronoid process, ramus and body</td>
<td>Numerous supernumerary teeth</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Borzabadi-Farahani et al.</td>
<td>F: 2 y</td>
<td>Mid-symphseal mandible</td>
<td>III</td>
<td>Anterior accessory mandible presenting as an oral mass</td>
<td>Twelve disorganized unerupted teeth and a dentigerous cyst</td>
<td>Submucosal cleft palate; cleft lower lip; oropharyngeal mass</td>
<td>Bimanual dyskinesis; webbing of the epicantal folds, scoliosis, ptosis, nasal choanal mass</td>
</tr>
<tr>
<td>Suhaili et al.</td>
<td>F: 4.5 m-3 y</td>
<td>Right mandibular body</td>
<td>III</td>
<td>Partial duplication of posterior alveolar process</td>
<td>Numerous supernumerary teeth</td>
<td>Duplicated lower lip; accessory mouth; macrostomia</td>
<td>No</td>
</tr>
<tr>
<td>Akpuaka and Nwozo</td>
<td>F: 6 m</td>
<td>Right mandibular body</td>
<td>III</td>
<td>Attached to the outer aspect of the right mandibular body</td>
<td>A number of irregularly arranged teeth</td>
<td>Accessory lower lip and macrostomia</td>
<td>No</td>
</tr>
<tr>
<td>Maisels</td>
<td>F: newborn to 17 y</td>
<td>Right mandibular body</td>
<td>III</td>
<td>Accessory mandible attached to the outer aspect of the right body of the true mandible, posterior to the mental foramen</td>
<td>A number of tooth follicles</td>
<td>Accessory mouth with a curtain of mucosa separating the 2 cavities, no accessory tongue</td>
<td>Split notochord syndrome</td>
</tr>
<tr>
<td>Price and Zarem</td>
<td>F: 6 y</td>
<td>Right mandible</td>
<td>III</td>
<td>Duplication of the right mandible</td>
<td>NS</td>
<td>Partial duplication of the mouth</td>
<td>No</td>
</tr>
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</table>

(continued on next page)
cleft and parotid aplasia can occur together with the duplication of the mandible.

REFERENCES


Table 1. Continued

<table>
<thead>
<tr>
<th>Author</th>
<th>Location</th>
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<th>Features</th>
<th>Supernumerary teeth</th>
<th>Other body site deformity (potential syndromes)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Beatty et al.</td>
<td>Right mandible</td>
<td>III</td>
<td>Circular alveolar process at the angle of the right mandible</td>
<td>Six supernumerary teeth like mandibular molars and canines</td>
<td>Transverse facial cleft; macrostomia</td>
</tr>
<tr>
<td>Lawrence et al.</td>
<td>Bilateral rami</td>
<td>IV</td>
<td>Bilateral accessory rami duplication</td>
<td>Toothy-bearing a premaxilar process</td>
<td>NS</td>
</tr>
<tr>
<td>Laws et al.</td>
<td>Bilateral rami</td>
<td>IV</td>
<td>Bilateral accessory rami duplication</td>
<td>Partly developed single-rooted tooth resembling a premolar</td>
<td>NS</td>
</tr>
<tr>
<td>Present study</td>
<td>Left mandibular body</td>
<td>III</td>
<td>Duplication of the posterior alveolar bone and rami duplication</td>
<td>Tooth-bearing characteristics</td>
<td>NS</td>
</tr>
</tbody>
</table>

F, female; M, male; y, year; m, month; NS, not specified.

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