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 arches[2] 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]

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 bold 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 2*B*). 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.

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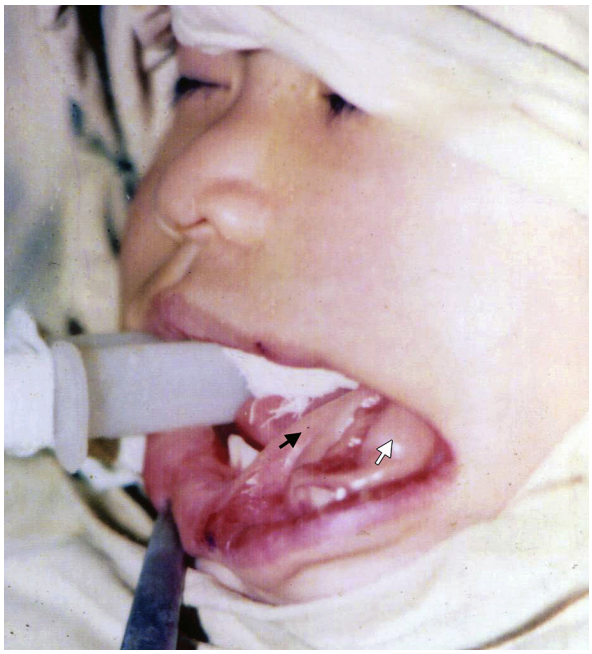

Fig. 1. Clinical photograph taken when the patient was 2 years old at the first surgery shows the left transverse facial cleft. An intra-oral soft tissue band (black arrow) divides the lower lip and the oral cavity into 2 parts. The accessory mandible is marked by white arrow.

trabecular bone visually and pathologically. Six supernumerary teeth were extracted (Figure 10*A*). Radiographs of the 3 molar-like teeth showed relatively normal morphology of the pulp chambers (Figure 10*B*). 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 dicephalus, diprosopus to partial facial duplication.[1,2] Wu et al. reviewed the literature and reported that approximately 100 cases of complete or incomplete craniofacial duplication were identified.[2]

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.[2,9] 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.[3-5] Type III is the alveolar type, characterized by the presence of a localized accessory alveolar bone with supernumerary teeth

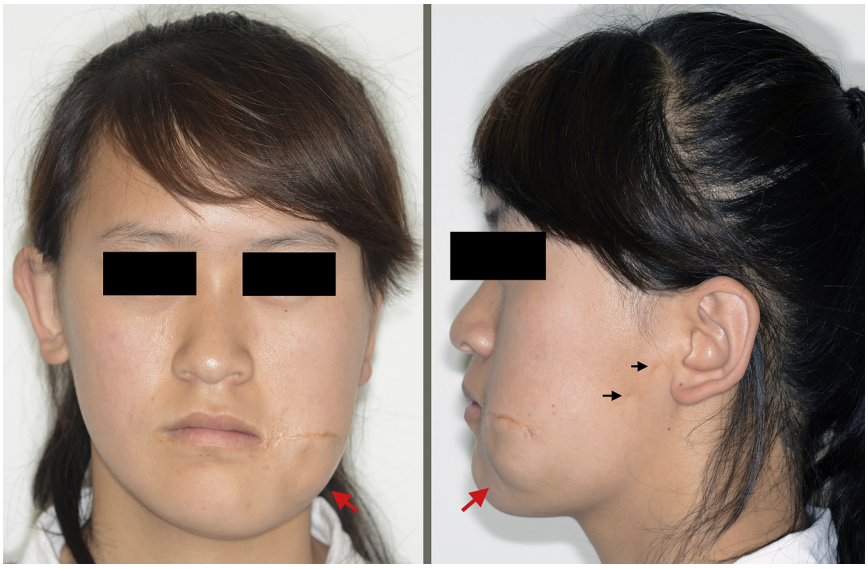

Fig. 2. Clinical photographs show the left facial scar due to surgical repair of the facial cleft. Two skin dimples (black arrows) are found in the left preauricular region. Note the bulging of the left mandibular facial area (red arrows).

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 attached to the normal mandible.[7,8,13] The mouth may be partially duplicated to present as macrostomia[8] or completely duplicated to present as a separate mouth[4] in the type II and III deformities. The supernumberary teeth in the duplicated mandible are frequently of regular morphology.[5] Type IV is characterized by the bilaterally

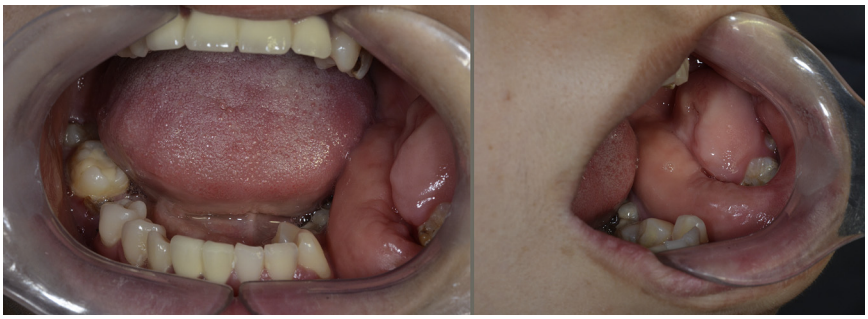

Fig. 3. Intra-oral views show that the permanent mandibular left molars and premolars are displaced. A soft tissue band is seen and reminiscent of the duplicated lower lips. A bony segment with 2 erupted supernumerary teeth and contact overlying mucosa locates anterior to the ramus.

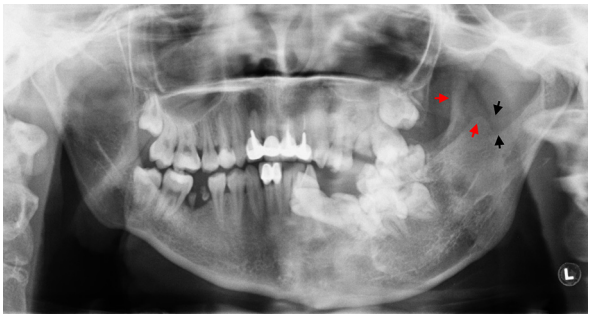

Fig. 4. Panoramic radiograph shows that the left mandibular ramus is duplicated with 2 coronoid processes (red arrows) and sigmoid notches (black arrows).

duplicated ramus and its remarkable association with Klippel–Feil syndrome.[10,12]

Facial cleft deformity is closely related to the duplicated mandible and maxilla.[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.[8,13]

Duplication of the ramus may be complete or partial in duplicated mandibles.[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,[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.[5] CT or MRI (magnetic resonance imaging)

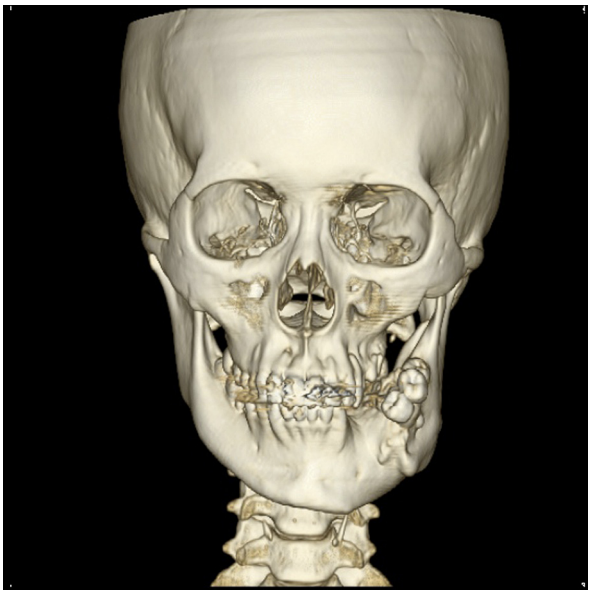

Fig. 5. Three-dimensional CT image shows that a redundant alveolar bone segment with a group of supernumerary teeth locates anterior to the ramus and lateral to the inherent left mandibular arch.

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.[24] Ectopic underdeveloped parotid gland can be observed superficial to the masseter.[24,25] Parotid aplasia in present case is quite similar to that described by Higley et al.[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.[26]

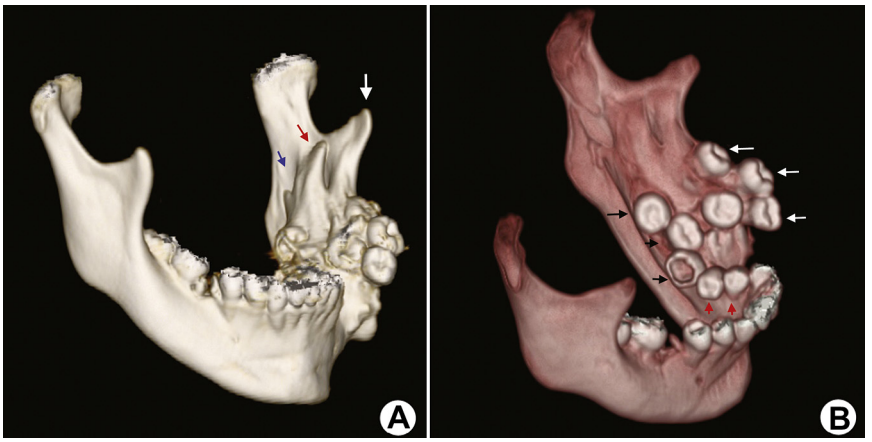

Fig. 6. Three-dimensional CT images of the mandible show that an outer greater coronoid process (white arrow, **A**) and an inner lesser coronoid process (red arrow, **A**) are present. Note the mandibular foramen (blue arrow, **A**) opening into the internal side of the ramus. The permanent mandibular 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.

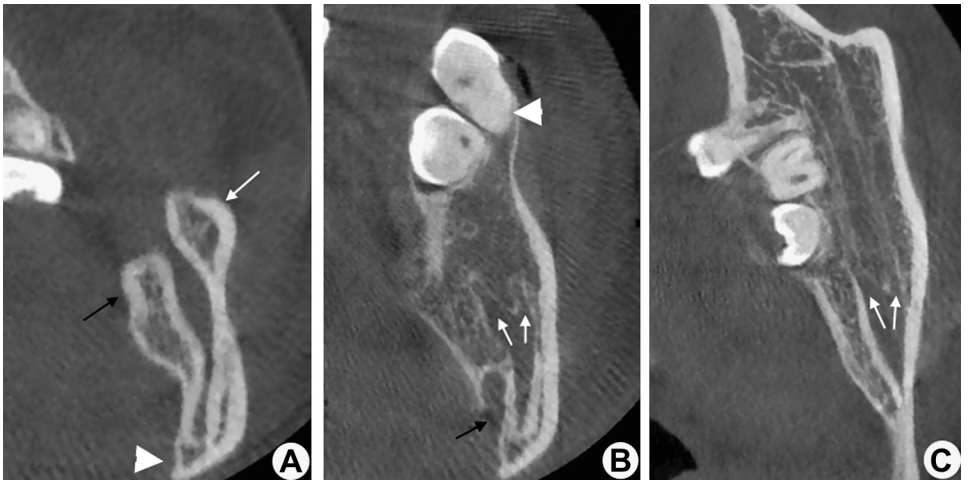

Fig. 7. Axial cone beam CT images (**A-B**) showing the structure of the duplicated ramus. The ramus is partially duplicated to form an outer (white arrow, **A**) and inner (black arrow, **A**) coronoid processes. The 2 processes fused together at the posterior border of the ramus (white arrow head, **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).

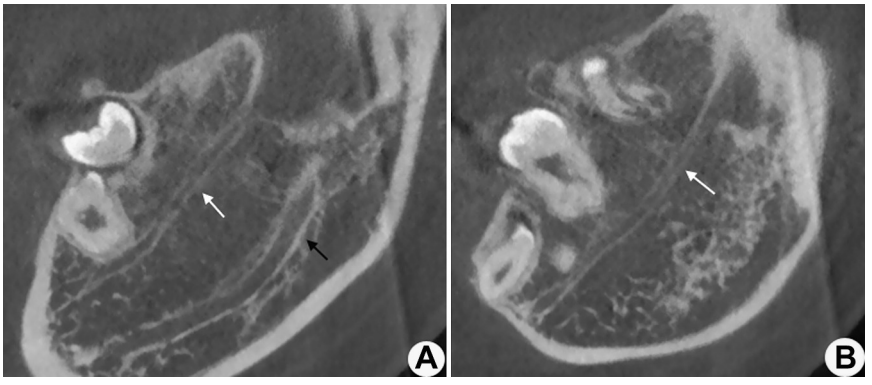

Fig. 8. Oblique reformatted cone beam CT images showing the inherent mandibular canal (black arrow, **A**) and redundant mandibular canals (white arrows, **A-B**).

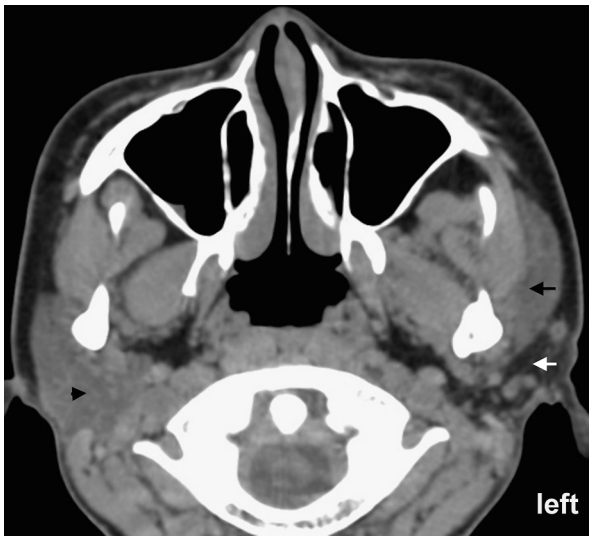

Fig. 9. Axial spiral CT image shows that the parotid gland is absent in the left parotid space (white arrow). Ectopic parotid gland tissue (black arrow) is observed superficial to the left masseter. Note the normal appearance of the right parotid gland (black arrow head).

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.

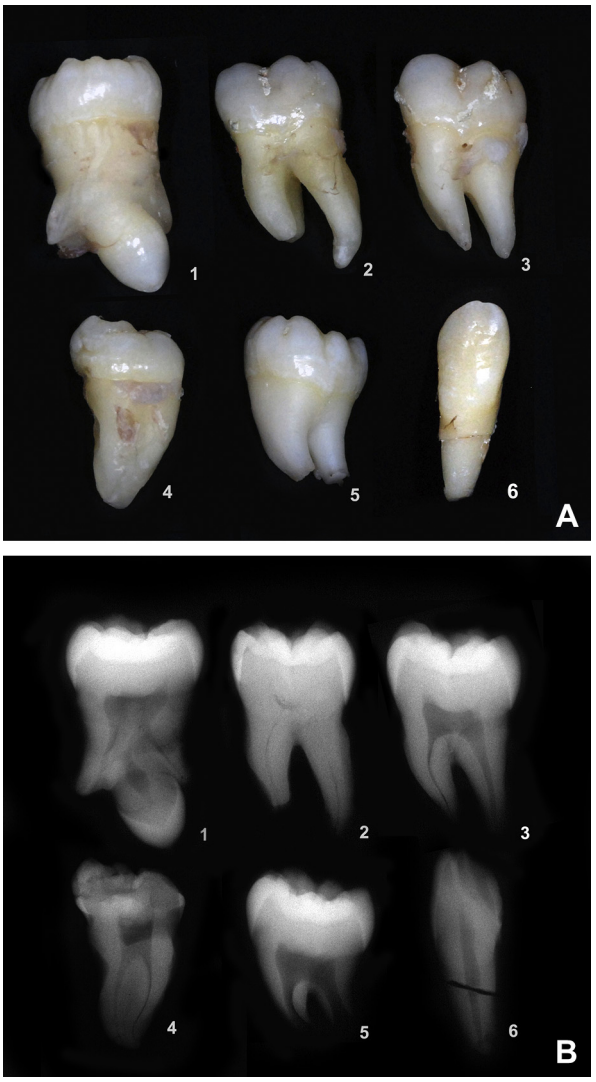

Fig. 10. The supernumerary teeth (**A**) in the accessory bone segment and their X-ray images (**B**). Tooth 1 is fused with another cone-shaped tooth at the roots. Narrowed pulp chamber and root canals are observed in tooth 2. Teeth 3-5 are of the regular molars shape. Tooth 6 is of mandibular canine shape. The crown of tooth 4 is partially destroyed and the root of tooth 6 fractured during the surgery.

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

Author	*Sex: age*	*Location*	*Type*	*Features*	*Supernumerary teeth*	*Other oral facial deformity*	*Other body site deformity (potential syndromes)*
Wittkampf and van Limborgh[9]	F: 3 y	Symmetric duplication	I	W-shaped double mandible with osseous connection in the midline	Multiple deciduous teeth in the mandibles	Hypertelorism, macrostomia, duplicate lower lip, cleft palate, duplicate anterior tongue	Intraoral hamartoma, duplication of sella turcica and odontoid processes, fused vertebrae C2 and C3; (split notochord syndrome)
Wu et al.[2]	M: newborn	Symmetric duplication	I	Two partially formed, separate mandibular arches	Teeth buds present in 2 mandibular arches	Duplicate tongue and upper labial frenulum; cleft palate	Exotropia; microphallus; generalized hypotonia; low-set ears; orbital hypertelorism; abnormal tectum; duplicate vertebral bodies from C2 to C5, C6 to T3
Mclaughlin[4]	F: 3 m	Right mandibular body	II	Duplicated horizontal rami fused with the normal mandible to the left of the symphysis	Accessory mandible contained 8 teeth buds	Duplication of mouth and tongue	NS
Davies et al.[5]	NS: 2 y	Right mandible	II	Accessory mandibular body fused with the enlarged ramus extending toward the temporomandibular joint	Eight supernumerary deciduous and permanent teeth with normal morphology	Duplicate mouth, accessory salivary gland duct	No
Shaikh et al.[3]	F: 30 y	Left mandibular body and ramus	II	Duplication of the hemi-mandible, condyle, coronoid process, ramus and body	Numerous supernumerary teeth	No	No
Borzabadi-Farahani et al.[14]	F: 2 y	Mid-symphyseal mandible	III	Anterior accessory mandible presenting as an oral mass	Twelve disorganized unerupted teeth and a dentigerous cyst	Submucosal cleft palate; cleft lower lip; oropharyngeal mass	Bimanual dyskinesis; webbing of the epicanthal folds, scoliosis, ptosis, nasal choanal mass
Suhaili et al.[13]	F: 4.5 m-3 y	Right mandibular body	III	Partial duplication of posterior alveolar process	Numerous supernumerary teeth	Duplicated lower lip; accessory mouth; macrostomia	No
Akpuaka and Nwozo[11]	F: 6 m	Right mandibular body	III	Attached to the outer aspect of the right mandibular body	A number of irregularly arranged teeth	Accessory lower lip and macrostomia	No
Maisels[8]	F: newborn to 17 y	Right mandibular body	III	Accessory mandible attached to the outer aspect of the right body of the true mandible, posterior to the mental foramen	A number of tooth follicles	Accessory mouth with a curtain of mucosa separating the 2 cavities, no accessory tongue	Split notochord syndrome
Price and Zarem[7]	F: 6 y	Right mandible	III	Duplication of the right mandible	NS	Partial duplication of the mouth	No

(*continued on next page*)

Table I. Continued

Author	Sex: age	Location	Type	Features	Supernumerary teeth	Other oral facial deformity	Other body site deformity (potential syndromes)
Beatty[6]	M: 3.5 y	Right mandible	III	Circular alveolar process at the angle of the right mandible	NS	Accessory mouth with well formed lips and a tongue at the angle of the right mandible	Right anophthalmia; left polycystic kidney
Al-Ani et al.[12]	M: 5 y	Bilateral rami	IV	Bilateral mandibular ramus duplication	Tooth-bearing	Submucous cleft palate	Occipital cleft; enlarged foramen magnum; (Klippel–Feil syndrome, Pierre Robin sequence)
Lawrence et al.[10]	M: 6 y	Bilateral rami	IV	Bilateral accessory rami	Partly developed single-rooted tooth resembling a premolar	Cleft palate; macrostomia; macroglossia and microgenia	Fusion of C3-C4 and C5-C6, short neck; thoracic kyphosis, ocular defect; hearing loss; ventricular septal defect; (Klippel–Feil syndrome)
Present study	F: 15 y	Left mandibular body	III	Duplication of the posterior alveolar bone and ramus	Six supernumerary teeth like mandibular molars and canines	Transverse facial cleft; macrostomia	NS

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

cleft and parotid aplasia can occur together with the duplication of the mandible.

REFERENCES

1. Farman AG, Escobar V. Duplication of oral and maxillofacial structures. *Quintessence Int.* 1986;17:731-737.
2. Wu J, Staffenberg DA, Mulliken JB, Shanske AL. Diprosopus: a unique case and review of the literature. *Teratology.* 2002;66: 282-287.
3. Shaikh MF, Naik N, Shah C. Duplication of hemi mandible and oral cavity, presentation of an adult patient—a case report. *J Plast Reconstr Aesthet Surg.* 2008;61:183-185.
4. McLaughlin CR. Reduplication of mouth, tongue, and mandible. *Br J Plast Surg.* 1948;1:89-95.
5. Davies D, Morrison G, Miller BH. Reduplication of the mouth and mandible. *Br J Plast Surg.* 1973;26:84-89.
6. Beatty HG. A report of a case of an unusual embryologic defect of the face. *Plast Reconstr Surg (1946).* 1956;17:297-303.
7. Price JE Jr, Zarem HA. Duplication of the mandible. *Plast Reconstr Surg.* 1979;64:104-105.
8. Maisels DO. Reduplication of the mouth and mandible. *Br J Plast Surg.* 1981;34:23-25.
9. Wittkampf AR, van Limborgh J. Duplication of structures around the stomatodeum. *J Maxillofac Surg.* 1984;12:17-20.
10. Lawrence TM, McClatchey KD, Fonseca RJ. Congenital duplication of mandibular rami in Klippel-Feil syndrome. *J Oral Med.* 1985;40:120-122.
11. Akpuaka FC, Nwozo JC. Reduplication of the mouth and mandible. *Plast Reconstr Surg.* 1990;86:971-972.
12. Al-Ani SA, Rees M, de Chalain TM. Our experiences managing a patient with mandibular duplication and cervical spinal fusion. *J Craniofac Surg.* 2009;20:2118-2122.
13. Suhaili DN, Somasundaram S, Lau SH, Ajura AJ, Roslan AR, Ramli R. Duplication of lower lip and mandible—a rare diprosopus. *Int J Pediatr Otorhinolaryngol.* 2011;75: 131-133.
14. Borzabadi-Farahani A, Gross J, Sanchez-Lara P, Yen SL. An unusual accessory mandible and a sub-mucosal cleft palate-a case report and review of the literature. *Cleft Palate Craniofac J.* March 2012. [Epub ahead of print].
15. Cameron AC, McKellar GM, Widmer RP. A case of neurocristopathy that manifests facial clefting and maxillary duplication. *Oral Surg Oral Med Oral Pathol.* 1993;75:338-342.
16. Borzabadi-Farahani A, Yen SL, Yamashita DD, Sanchez-Lara PA. Bilateral maxillary duplication: case report and literature review. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2012;113: e29-e32.
17. Tharanon W, Ellis E 3rd, Sinn DP. A case of maxillary and zygomatic duplication. *J Oral Maxillofac Surg.* 1998;56: 770-774.
18. Cheung LK, Samman N, Tideman H. Bilateral transverse facial clefts and accessory maxillae—variant or separate entity? *J Craniomaxillofac Surg.* 1993;21:163-167.
19. Sjamsudin J, David D, Singh GD. An Indonesian child with orofacial duplication and neurocristopathy anomalies: case report. *J Craniomaxillofac Surg.* 2001;29:195-197.
20. Jian XC, Chen XQ, Hunan C. Neurocristopathy that manifests right facial cleft and right maxillary duplication. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 1995;79: 546-550.
21. Miyajima K, Natsume N, Kawai T, Iizuka T. Oblique facial cleft, cleft palate, and supernumerary teeth secondary to amniotic bands. *Cleft Palate Craniofac J.* 1994;31:483-486.
22. Smylski PT. Accessory jaw bones; report of case. *J Oral Surg (Chic).* 1952;10:70-74.

23. Rushton MA, Walker FA. Unilateral secondary facial cleft with excess tooth and bone formation. *Proc R Soc Med.* 1936;30:79-82.
24. Higley MJ, Walkiewicz TW, Miller JH, Curran JG, Towbin RB. Aplasia of the parotid glands with accessory parotid tissue. *Pediatr Radiol.* 2010;40:345-347.
25. Antoniades DZ, Markopoulos AK, Deligianni E, Andreadis D. Bilateral aplasia of parotid glands correlated with accessory parotid tissue. *J Laryngol Otol.* 2006;120:327-329.
26. Barr M Jr. Facial duplication: case, review, and embryogenesis. *Teratology.* 1982;25:153-159.
27. Muraskas JK, McDonnell JF, Chudik RJ, Salyer KE, Glynn L. Amniotic band syndrome with significant orofacial clefts and disruptions and distortions of craniofacial structures. *J Pediatr Surg.* 2003;38:635-638.

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