A rare case of accessory maxilla: a case report and literature review of Tessier no. 7 clefts

Ming Sun1, Na Lv1, Ya Xiao1, Jiabin Li2,3,4,5 and Guangzhao Guan6

Abstract
Bilateral Tessier no. 7 clefts are rarely reported in the literature. Here, we describe the presence of accessory maxilla with supernumerary teeth in a patient who exhibited bilateral Tessier no. 7 clefts; the diagnosis was established based on the patient's history, clinical presentation, and computed tomography images. A review of the available literature revealed 24 patients with Tessier no. 7 clefts from 2000 to 2020, including our patient. The most common clinical manifestation in patients with Tessier no. 7 clefts comprises bilateral facial clefts. Additionally, Tessier no. 7 clefts are more frequently found in boys or men, rather than in girls or women. The presence of an accessory maxilla with supernumerary teeth in a patient with bilateral Tessier no. 7 clefts is extremely rare. Early detection of craniofacial abnormalities is important, because it may influence patient prognosis and management.

Keywords
Macrostomia, bilateral Tessier no. 7 clefts, accessory maxilla, supernumerary teeth, craniofacial abnormalities, facial clefts, Tessier clefting system

Date received: 22 December 2019; accepted: 20 April 2020

1Department of Stomatology, the First Affiliated Hospital of Anhui Medical University, Hefei, China
2Department of Infectious Diseases, the First Affiliated Hospital of Anhui Medical University, Hefei, China
3Anhui Center for Surveillance of Bacterial Resistance, Hefei, China
4Institute of Bacterial Resistance, Anhui Medical University, Hefei, China
5Department of Infectious Diseases, the Chaohu Affiliated Hospital of Anhui Medical University, Hefei, China
6Department of Oral Diagnostic and Surgical Sciences, Faculty of Dentistry, University of Otago, Dunedin, New Zealand

Corresponding author:
Guangzhao Guan, Department of Oral Diagnostic and Surgical Sciences, Faculty of Dentistry, University of Otago, 310 Great King Street, Dunedin, 9016, New Zealand.
Email: simon.guan@otago.ac.nz
Introduction

Craniofacial clefts are uncommon congenital deformities. The etiology of craniofacial clefts is unknown; they presumably occur as a result of genetic predisposition and environmental factors (e.g., infection, medication, and radiation). The exact incidence is unclear, although craniofacial clefts are estimated to be present in 1.4 to 4.9 of 100,000 births. Currently, there is no universal classification for orofacial and craniofacial clefts. The Tessier clefting system is a commonly used classification approach, based on anatomic and descriptive features. This classification assigns numbers (1–30) to various sites of clefting, depending on their anatomic relationship with the sagittal midline. Tessier no. 0 (59.5%) is the most common type of craniofacial cleft, whereas Tessier no. 8, 13, and 30 are the rarest types of clefts.

Tessier no. 7 clefts are uncommon, comprising 5.5% to 13.9% of all clefts. They are characterized by macrostomia, facial muscular diastasis, and abnormalities of the maxilla and zygomatic bone. The exact etiology of these clefts has not yet been determined; they may arise from failed fusion of the embryonic mandibular and maxillary process at the first pharyngeal arch. Tessier no. 7 clefts might be associated with other anomalies, such as an accessory maxilla or mandible. The terms “accessory maxilla” and “maxillary duplication” refer to a rare clinical entity that is characterized by the presence of extra bone, lying posterior to the maxillary tuberosity. The accessory maxilla is presumably caused by abnormal growth of the zygomatic arch and is often associated with facial clefts, including Tessier no. 7 clefts. However, the presence of an accessory maxilla in a patient with a Tessier no. 7 cleft is extremely rare. To the best of our knowledge, there have been four reported cases in the literature. Here, we describe the presence of an accessory maxilla in a 25-year-old man with bilateral Tessier no. 7 clefts.

Case report

In March 2018, a 25-year-old man with a history of bilateral Tessier no. 7 clefts was referred to the First Affiliated Hospital of Anhui Medical University for evaluation and treatment of an enlarged asymptomatic mass involving supernumerary teeth on the right posterior maxilla, which he had noticed 10 years prior to referral. The patient’s medical history included surgical repair of bilateral Tessier no. 7 clefts when he was 15 years old. He did not take any regular medications and supplements; additionally, he did not smoke and rarely drank alcohol. He reported no family history of facial deformities or inherited diseases.

Extraoral examination revealed no regional lymphadenopathy or swollen salivary glands. The patient’s right zygomatic bone was more prominent than his left zygomatic bone. Although surgical scars were noted at both commissures, the patient’s mouth opening was not limited by the presence of scar tissue (Figure 1). He exhibited no abnormalities in the external and middle ears.

Figure 1. Clinical photo showing facial asymmetry.
Intraoral examination revealed an outgrowth of bone with supernumerary teeth on the right posterior maxilla (Figure 2). With the exception of two narrow mucosal grooves on the right posterior maxilla, no alveolar cleft was observed. Teeth 13 and 24 were absent. The remaining oral structures were normal.

Panoramic radiography and computed tomography scans of the patient’s cranio-maxillofacial skeleton also revealed an outgrowth of bone with supernumerary teeth, which appeared to arise from the right posterior maxilla; this outgrowth extended from the inferior border of the right zygomatic bone to the maxillary tuberosity. In addition, a gap was observed between the normal maxilla and the outgrowth of bone on the right side (Figure 3). Based upon the patient’s history, clinical findings, and computed tomography findings, a diagnosis of accessory maxilla with bilateral Tessier no. 7 clefts was made.

Surgical reconstruction was performed with the patient under general anesthesia. An angular incision was made and a mucoperiosteal flap was raised. Massive sclerotic bone was found between the posterior maxilla and the zygomatic bone. The accessory maxillary region was divided sharply along the oblique connecting line of the posterior maxilla and zygomatic bone. Massive bone was removed from the right posterior maxilla. A gap was observed between the normal maxilla and the accessory maxilla (Figure 2). Massive bone was removed between the zygomatic bone and the maxillary tuberosity. The posterior maxilla and zygomatic bone were divided into two pieces along the zygomatic arch. Sclerotic bone was found along the posterior maxilla and zygomatic bone. All bone fragments were removed and the surgical field was irrigated with saline. The bone was repositioned and rigidly fixed with miniplates (Figure 3). The surgical wound was irrigated with saline and closed with 3-0 Vicryl sutures. The surgical wound was covered with a sterile dressing. The patient had a healthy oral cavity postoperatively.

Figure 2. Intraoral photo showing right posterior accessory maxilla with supernumerary teeth (black arrows: two mucosal grooves; red arrow: supernumerary teeth).

Figure 3. Preoperative computed tomography images. (a) Three-dimensional reconstruction computed tomography scan (black arrow: bony segments extending from inferior borders of right zygomatic bone to maxillary tuberosity; red arrow: bony gap between normal maxilla and teeth-bearing bony segments on right side). (b) Axial view of computed tomography scan (red arrow: high-density mass at right posterior maxillary region).
maxilla with supernumerary teeth was surgically removed; the excised specimen measured 4.5 cm × 3.0 cm × 2.0 cm (Figure 4). No bone graft was required, and the maxillary sinus remained intact during the surgery. Subsequent histopathological analysis revealed tooth-like structures, osseous tissue, and fibrous connective tissue in the excised specimen (Figure 5); no abnormal cells were found. By the 3-month follow-up examination, the wound had healed completely (Figure 6). The patient did not report any postoperative complications. Both postoperative computed tomography and panoramic radiography scans showed no signs of local recurrence (Figure 7).

Ethical approval was obtained from the local ethics committee (approval no. 20190127) for treatment and publication of this report. Written consent was obtained from the patient for treatment and publication of this report.

**Discussion**

It is important to examine fetal craniofacial structure during prenatal examinations because abnormalities involving these
structures may suggest the presence of syndromes, chromosomal abnormalities, infectious diseases, or metabolic disorders. Prenatal ultrasounds (e.g., two-dimensional, three-dimensional, or four-dimensional ultrasound) are accurate, reliable, and non-invasive examination tools that can be used for detection of craniofacial abnormalities. Current international guidelines recommend routine mid-trimester ultrasound scans for evaluation of the fetal face including the upper lip, median facial profile, orbits, nose, and nostrils.

This report described a man with bilateral Tessier no. 7 clefts who presented for treatment of a right accessory maxilla. Thus far, the etiology of Tessier no. 7 clefts is unclear; it may involve amniotic membrane syndrome, failed maxillary fusion of the first branchial arch, or failed mesodermal migration. Hard and soft tissue deformities are present in patients with Tessier no. 7 facial clefts. The hard tissue abnormalities are characterized by deformation of the cranial base, glenoid fossa, sphenoid, condyle, coronoid process, mandibular ramus, posterior maxilla, alveolar process, and alveolar process, as well as the absence of zygomatic arch and maxillary cleft in the molar region; deformation of the maxillary tuberosity and pterygoid process are also observed. The soft tissue abnormalities are characterized by macrostomia, external and middle ear abnormalities, and temporalis abnormality, as well as sporadic abnormalities of the parotid gland and cranial nerves 5 and 7.

In the available literature, 24 patients with Tessier no. 7 clefts have been described from 2000 to 2020, including our patient. The clinical findings of these patients are presented in Table 1. Among the 24 patients, 12 were male patients and eight were female patients; sex could not be identified in four patients. The most common clinical manifestation of Tessier no. 7 clefts was bilateral facial clefts, which was present in 66.7% of the patients. Syndromes (i.e., Goldenhar Syndrome, Dandy-Walker Syndrome, Amniotic band syndrome, and posterior fossa brain malformations, hemangioma, arterial lesions,
| Authors                      | Year of publication | Sex | Age               | Bilateral/unilateral facial cleft | Location of duplication | Normal teeth | Supernumerary teeth | Mandibular dysplasia | Alveolar cleft | Syndromes                     |
|------------------------------|---------------------|-----|-------------------|-----------------------------------|-------------------------|--------------|---------------------|---------------------|---------------|--------------------------------|
| Witters et al.               | 2001                | M   | 37 weeks of gestation | Unilateral left                     | N/A                     | N/A          | N/A                 | N/A                 | N/A           | Goldenhar Syndrome             |
| Shima et al.                 | 2002                | F   | 35 weeks of gestation | Bilateral                          | N/A                     | N/A          | N/A                 | N/A                 | N/A           |                                |
| Presti et al.                | 2004                | N/A | 1 year            | Bilateral                          | N/A                     | N/A          | N/A                 | N/A                 | N/A           |                                |
| Sigler et al.                | 2004                | M   | 6 months          | Unilateral left                     | N/A                     | N/A          | N/A                 | N/A                 | N/A           |                                |
| Pilu et al.                  | 2005                | N/A | 22 weeks of gestation | Bilateral                          | N/A                     | N/A          | N/A                 | N/A                 | N/A           |                                |
| Bodin et al.                 | 2006                | F   | 4 months           | Bilateral                          | N/A                     | N/A          | N/A                 | Yes                 | Yes           | N/A                            |
| Kuriyama et al.              | 2008                | M   | 1 year            | Bilateral                          | N/A                     | N/A          | Yes                 | Yes                 | N/A           | N/A                            |
| Dasgupta et al.              | 2009                | M   | 5 years           | Bilateral                          | N/A                     | N/A          | Yes                 | Yes                 | N/A           | N/A                            |
| Asai et al.                  | 2010                | F   | 1 year            | Bilateral                          | N/A                     | N/A          | Yes                 | Yes                 | N/A           | N/A                            |
| Ahmed et al.                 | 2010                | M   | 7 years           | Bilateral                          | N/A                     | N/A          | Yes                 | N/A                 | N/A           | N/A                            |
| Bajaj et al.                 | 2011                | M   | 7 months          | Unilateral left                     | N/A                     | Yes          | N/A                 | Yes                 | N/A           | PHACE syndrome                  |
| Troyano Luque et al.         | 2011                | N/A | 1 year            | Unilateral left                     | N/A                     | N/A          | N/A                 | N/A                 | N/A           | N/A                            |
| Hou et al.                   | 2011                | M   | 3 years           | Bilateral                          | N/A                     | Yes          | N/A                 | Yes                 | Yes           | N/A                            |
| Uchikawa et al.              | 2011                | F   | 2 years           | Bilateral                          | N/A                     | N/A          | N/A                 | Yes                 | N/A           | N/A                            |
| Dhupar et al.                | 2012                | M   | 6 months          | Unilateral right                    | N/A                     | Yes          | N/A                 | N/A                 | N/A           | Dandy-Walker Syndrome           |
| Chang et al.                 | 2012                | M   | 40 weeks of gestation | Unilateral right                    | N/A                     | N/A          | N/A                 | N/A                 | N/A           |                                |
| Borzabadi-Farahani et al.    | 2013                | M   | 13 years          | Bilateral                          | Right accessory maxilla  | Yes          | Yes                 | Yes                 | Yes           | N/A                            |
| Ozçelik et al.               | 2014                | F   | 50 days           | Unilateral right                    | Right accessory mandible | N/A          | N/A                 | N/A                 | N/A           | N/A                            |
| Chauhan and Guruprasad       | 2015                | F   | 11 days           | Bilateral                          | Left accessory maxilla   | Yes          | Yes                 | Yes                 | Yes           | Goldenhar Syndrome             |
| Chauhan and Guruprasad       | 2015                | M   | 18 years          | Bilateral                          | Left and right accessory maxilla | Yes       | Yes                 | N/A                 | Yes           | N/A                            |
| Yin et al.                   | 2017                | F   | 10 years          | Bilateral                          | Left and right accessory maxilla | Yes       | Yes                 | N/A                 | N/A           | Amniotic band syndrome          |
| Cavaco-Gomes et al.          | 2017                | N/A | 23 weeks and 5 days of gestation | Unilateral left                    | N/A                     | N/A          | N/A                 | N/A                 | N/A           | N/A                            |
| Raveendran et al.            | 2018                | F   | 1 year            | Bilateral                          | N/A                     | N/A          | N/A                 | N/A                 | N/A           |                                |
| This report                  | 2020                | M   | 25 years          | Bilateral                          | Right accessory maxilla  | Yes          | Yes                 | Yes                 | Yes           | N/A                            |

Abbreviations: F, female; M, male; N/A, information not available; PHACE, posterior fossa brain malformations, hemangioma, arterial lesions, cardiac abnormalities, and eye abnormalities.
cardiac abnormalities, and eye abnormalities (PHACE syndrome) were present in five patients. Three patients exhibited a unilateral accessory maxilla, two exhibited bilateral accessory maxillae, and one exhibited a right accessory mandible. Supernumerary teeth were present in five patients with jaw duplication; alveolar cleft and mandibular dysplasia were present in seven patients. All patients were described in case reports. Most patients had a prenatal diagnosis and had undergone reconstructive surgical treatment.

Duplication of jaws with supernumerary teeth can occur in maxilla or mandible, either as an accessory outgrowth or a complete jaw. The exact incidence of jaw duplication is unclear; it is estimated that 1 in 80,000 births in the general population may be affected, with a higher incidence in boys than in girls and a higher incidence in the maxilla than in the mandible. The majority of published reports described unilateral involvement; fewer than 20% of patients exhibited bilateral involvement. Maxillary duplication is a rare congenital condition that is often accompanied by cleft lip, palate, or other craniofacial clefts.

To the best of our knowledge, only a few instances of maxillary duplication have been described in the literature. Accessory maxilla as a type of maxillary duplication has often been reported with facial clefts; isolated maxillary duplication with no other anomalies has also been described.

Early detection of jaw duplication is important because it could influence the growth of the craniomaxillofacial region (e.g., tooth eruption and facial growth). Thus, we recommend early referral to tertiary centers and regular follow-up of affected patients to rule out the presence of other craniofacial anomalies and syndromes.

Conclusion

The most common clinical manifestation in patients with Tessier no. 7 clefts comprises bilateral facial clefts. Additionally, Tessier no. 7 clefts are more frequently found in boys or men, rather than in girls or women. The presence of an accessory maxilla with supernumerary teeth in a patient with bilateral Tessier no. 7 clefts is rare. This report emphasizes that early detection of craniofacial abnormalities is important, because it may influence patient prognosis and management.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Funding

We thank Anhui Provincial Higher Education Talent Project, gxyq 2018010 and Natural Science Research Project of Anhui Higher Education Institutions, KJ2018A0193, for the support of the publication cost.

ORCID iD

Guangzhao Guan https://orcid.org/0000-0001-7265-9865

References

1. Kalantar-Hormozi A, Abbaszadeh-Kasbi A, Goravanchi F, et al. Prevalence of rare craniofacial clefts. J Craniofac Surg 2017; 28: e467–e470.
2. Kawamoto HK Jr. The kaleidoscopic world of rare craniofacial clefts: order out of chaos (Tessier classification). Clin Plast Surg 1976; 3: 529–572.
3. Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. J Maxillofac Surg 1976; 4: 69–92.
4. Fearon JA. Rare craniofacial clefts: a surgical classification. J Craniofac Surg 2008; 19: 110–112.
5. Gorlin RJ. Syndromes of the head and neck. 4th ed. Oxford, England: Oxford University Press, 2001.
6. Woods RH, Varma S and David DJ. Tessier no. 7 cleft: a new subclassification and
management protocol. *Plast Reconstr Surg* 2008; 122: 898–905.

7. Chauhan DS and Guruprasad Y. Bilateral Tessier’s 7 cleft with maxillary duplication. *J Maxillofac Oral Surg* 2015; 14: 108–112.

8. Borzabadi-Farahani A, Yen SL, Yamashita DD, et al. Bilateral maxillary duplication: case report and literature review. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2012; 113: e29–e32.

9. Chauhan DS and Guruprasad Y. Goldenhar syndrome with Tessier’s 7 cleft: report of a case. *J Maxillofac Oral Surg* 2015; 14: 42–46.

10. Yin S, Ma RH, Li J, et al. A rare case of two accessory maxillae with bilateral Tessier 7 clefts. *Chin J Dent Res* 2017; 20: 53–58.

11. Mak ASL and Leung KY. Prenatal ultrasonography of craniofacial abnormalities. *Ultrasoundography (Seoul, Korea)* 2019; 38: 13–24.

12. Salomon LJ, Alfirevic Z, Berghella V, et al. Practice guidelines for performance of the routine mid-trimester fetal ultrasound scan. *Ultrasound Obstet Gynecol* 2011; 37: 116–126.

13. Borzabadi-Farahani A, Yen SL, Francis C, et al. A rare case of accessory maxilla and bilateral Tessier no. 7 clefts, a 10-year follow-up. *J Craniomaxillofac Surg* 2013; 41: 527–531.

14. Witters I, Schreurs J, Van Wing J, et al. Prenatal diagnosis of facial clefting as part of the oculo-auriculo-vertebral spectrum. *Prenat Diagn* 2001; 21: 62–64.

15. Shima Y, Ogawa K, Kuwabara Y, et al. Newborn with transverse facial cleft associated with polyhydramnios. *J Perinatol* 2002; 22: 91–92.

16. Presti F, Celentano C, Marcazzo L, et al. Ultrasound prenatal diagnosis of a lateral facial cleft (Tessier number 7). *Ultrasound Obstet Gynecol* 2004; 23: 606–608.

17. Sigler MO, Stein J and Zuker R. A rare craniofacial cleft: numbers 7, 2, and 3 clefts accompanied by a single median lip pit. *Cleft Palate Craniofac J* 2004; 41: 327–331.

18. Phlu G, Visentin A, Ambrosini G, et al. Three-dimensional sonography of unilateral Tessier number 7 cleft in a mid-trimester fetus. *Ultrasound Obstet Gynecol* 2005; 26: 98–99.

19. Bodin F, Salazard B, Bardot J, et al. Craniofacial cleft: a case of Tessier no. 3, 7 and 11 cleft. *J Plast Reconstr Aesthet Surg* 2006; 59: 1388–1390.

20. Kuriyama M, Udagawa A, Yoshimoto S, et al. Tessier number 7 cleft with oblique clefts of bilateral soft palates and rare symmetric structure of zygomatic arch. *J Plast Reconstr Aesthet Surg* 2008; 61: 447–450.

21. Dasgupta D, Jain A, Baxi V, et al. Fiberoptic intubation using LMA as a conduit and cook airway catheter as an exchanger in a case of Tessier 7 facial cleft syndrome. *Indian J Anaesth* 2009; 53: 230–232.

22. Asai S, Tanaka M, Miyakoshi K, et al. A case of Tessier number 7 cleft with severe micrognathia: prenatal sonographic and three-dimensional helical computed tomographic images. *Prenat Diagn* 2010; 30: 159–161.

23. Ahmed SS, Bey A, Hashmi SH, et al. Bilateral transverse facial cleft as an isolated and asyndromic deformity. *Int J Clin Pediatr Dent* 2010; 3: 101–104.

24. Bajaj A, Dyke P, Zaleski C, et al. Mild Tessier No. 7 cleft with PHACE syndrome: the case for pulmonary vascular steal. *Am J Med Genet A* 2011; 155a: 2298–2301.

25. Troyano Luque JM, Padilla Perez AI, Guerra Martin AL, et al. A case of isolated Tessier 7 cleft in the newborn of a diabetic mother. *J Obstet Gynaecol* 2011; 31: 343–345.

26. Hou R, Feng X, Zhang J, et al. A rare bilateral Tesser no. 6 and 7 clefts. *J Craniomaxillofac Surg* 2011; 39: 93–95.

27. Uchikawa Y, Ogata H, Hattori N, et al. Palatoplasty for bilateral oblique cleft of the soft palate accompanied by Tessier number 5.7 cleft. *Cleft Palate Craniofac J* 2011; 48: 231–235.

28. Dhupar V, Kumar P, Akkara F, et al. Dandy Walker syndrome with Tessier 7 cleft—a rare case report and a surgical note on the use of vermilion flap and lazy W-plasty. *J Maxillofac Oral Surg* 2012; 11: 368–370.
29. Chang YL, Lien R, Chang SD, et al. Prenatal 3D sonographic diagnosis of an isolated lateral facial cleft. *J Clin Ultrasound* 2012; 40: 219–221.

30. Ozcelik D, Toplu G, Turkseven A, et al. Lateral facial cleft associated with accessory mandible having teeth, absent parotid gland and peripheral facial weakness. *J Craniomaxillofac Surg* 2014; 42: e239–e244.

31. Cavaco-Gomes J, Duarte C, Pereira E, et al. Prenatal ultrasound diagnosis of Tessier number 7 cleft: case report and review of the literature. *J Obstet Gynaecol* 2017; 37: 421–427.

32. Raveendran JA, Chao JW, Rogers GF, et al. The “double” Tessier 7 cleft: an unusual presentation of a transverse facial cleft. *Cleft Palate Craniofac J* 2018; 55: 903–907.

33. Sun L, Sun Z and Ma X. Partial duplication of the mandible, parotid aplasia and facial cleft: a rare developmental disorder. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2013; 116: e202–e209.

34. Jian XC, Chen XQ and Hunan C. Neurocristopathy that manifests right facial cleft and right maxillary duplication. *Oral Surg Oral Med Oral Pathol Oral Radiol* 1995; 79: 546–550.

35. Morita K, Iwasa T, Imaiuzumi F, et al. A case of maxillary duplication with a soft palate reconstruction using a forearm flap. *Int J Oral Maxillofac Surg* 2008; 37: 862–865.

36. Cameron AC, McKellar GM and Widmer RP. A case of neurocristopathy that manifests facial clefting and maxillary duplication. *Oral Surg Oral Med Oral Pathol Oral Radiol* 1993; 75: 338–342.

37. Mafeni JO. The Robin sequence associated with partial maxillary duplication and multiple facial clefts: a case report. *Int J Oral Maxillofac Surg* 1993; 7: 31–34.

38. Kainulainen VT, Sandor GK and Stoneman DW. Management of a patient with an accessory maxilla and congenital facial fistula. *J Can Dent Assoc* 2005; 71: 161–163.

39. DeGurse K, Chung H and Pharoah M. Facial dimple with accessory bone and teeth. *Dentomaxillofac Radiol* 1995; 24: 135–138.

40. Hou M, Liu C, Wang J, et al. Lateral or oblique facial clefts associated with accessory maxillae: review of the literature and report of a case. *J Craniomaxillofac Surg* 2015; 43: 585–592.

41. Sun L, Sun Z and Ma X. Congenital parotid ectopia in accessory maxilla and facial cleft anomalies: three cases report. *Int J Pediatr Otorhinolaryngol* 2013; 77: 608–612.