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Cheek Fistula From the Ectopic Salivary Gland: A Variant of the Oculo-Auriculo-Vertebral Spectrum

Zhi-Peng Sun, MD, DDS; Xia Hong, DDS; Xu-Chen Ma, PhD, DDS; Zu-Yan Zhang, DDS, PhD; Guang-Yan Yu, PhD, DDS

Objectives/Hypothesis: This study aimed to investigate the diagnosis and management of a distinct developmental deformity syndrome characterized by congenital cheek fistula, ectopic accessory parotid gland, and preauricular appendage.

Study Design: Retrospective study.

Methods: We analyzed the medical records, radiologic and histopathologic findings, and follow-up data for seven patients (four males) with a congenital cheek salivary fistula. Computed tomography, fistulography, and sialography had been performed for diagnosis. Surgical treatment effect was evaluated.

Results: The mean age of the patients was 8.1 years (range, 2–16 years). The distinctive clinical feature was a congenital skin orifice lateral to the commissure with saliva discharge during eating. The cheek fistulae were accompanied by ipsilateral preauricular appendages in all seven patients. The skin orifice connected to an ectopic gland anterior to the masseter and inferior to Stensen's duct. Parotid sialography demonstrated an intact Stensen's duct in all cases. Hypoplasia of the ipsilateral mandible could be observed in five cases. Excision of the ectopic gland, skin orifice, and fistula was performed in five cases resulting in optimal treatment outcomes with no recurrent or adverse events.

Conclusions: A congenital saliva-discharging fistula with an ectopic accessory parotid gland, ipsilateral preauricular appendage, and mandibular hypoplasia constitutes a rare developmental syndrome. Surgical excision can effectively treat congenital cheek salivary fistula.

Key Words: Congenital, salivary gland, fistula, computed tomography, oculo-auriculo-vertebral spectrum.

Level of Evidence: 4.

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INTRODUCTION

The development of a congenital cheek salivary fistula (CCSF) from ectopic accessory gland tissue has been documented as a very rare clinical entity.[1–3] It is characterized by the occurrence of a saliva-discharging skin fistula lateral to the oral commissure.[2–9] An ipsilateral preauricular appendage is another frequent diagnostic sign. In our previous study, we have additionally reported that moderate mandibular hypoplasia constitutes another clinical feature.[2]

The oculo-auriculo-vertebral spectrum, also known as hemifacial microsomia, Goldenhar syndrome, and first and second branchial arch syndrome, embodies a wide range of hypoplastic malformations involving the temporal bone, outer ear, parotid gland, masseter, and mandible.[10,11] A preauricular appendage has been regarded as a very characteristic and mandatory facial feature of this spectrum.[11] It is reasonable to postulate that CCSF develops from disorders of the formation of the first branchial apparatus and constitutes a variant of this spectrum. However, the developmental embryogenesis is not clear.

Through further investigation of the clinical manifestations of this rarity and its rational treatment protocol, we aimed to further document this syndrome and to suggest a possible developmental mechanism and reasonable surgical protocol.

MATERIALS AND METHODS

From October 2008 to August 2013, seven patients affected with CCSF were referred to the Center of Salivary Gland Disease, Peking University School and Hospital of Stomatology. Their medical records, radiologic and histopathologic findings, and follow-up records were retrospectively reviewed. This study was approved by the institutional review board of Peking University School and Hospital of Stomatology (PKUSSIRB-2012084).

Noncontrast computed tomography (CT), CT or x-ray parotid sialography and fistulography had been used for radiologic evaluation. Noncontrast CT scans without the use of contrast medium were performed before fistulography and sialography. The fistulography was performed using CT (in six patients) and x-ray (in one patient) before sialography. For fistulography, contrast medium (1 mL, Iopamidol; Bracco, Milan, Italy) was injected via the cheek skin fistula. For sialography, contrast medium (1 mL) was injected via Stensen's duct.

From the Department of Oral and Maxillofacial Radiology (Z.-P.S., X.C.M., Z.-Y.Z.) and the Department of Oral and Maxillofacial Surgery (X.H., G.-Y.Y.), Peking University School and Hospital of Stomatology, Beijing, China.

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Send correspondence to Guang-Yan Yu, PhD, DDS, Department of Oral and Maxillofacial Surgery, Peking University School and Hospital of Stomatology, #22 Zhongguancun South Street, Haidian District, Beijing 100081, P. R. China. E-mail: gyyu@263.net

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TABLE I.
Clinical Information for the Seven Patients With Congenital Cheek Salivary Fistulae.

Patient No.	Gender	Age, yr	Side	Preauricular ppendage	Infection	Mandibular symmetry	Management	Follow-up Course (mo)
1	M	2	Left	Yes	No	Not noticed	Surgical excision	12
2	F	2	Right	Excised	Yes	Not noticed	Surgical excision	6
3	M	5	Left	Yes	No	Yes	Keep in follow-up	36
4	M	7	Right	Yes	No	Yes	Surgical excision	4
5	F	12	Right	Yes	No	Yes	Keep in follow-up	40
6	M	16	Left	Excised	No	Yes	Surgical excision	47
7	F	13	Left	Yes	No	Yes	Surgical excision	72

F = female; M = male.

The CT scans were initiated immediately after injection for CT sialography and fistulography. Spiral CT scans were performed using the GE BrightSpeed CT system (GE Healthcare, Chalfont St. Giles, United Kingdom) with the following parameters: 1.25 mm collimation, 16.75 mm/rotation table speed, 1:1.675 pitch, 80 to 120 kV, automatic exposure control. Axial images of 1.25 mm thickness were reconstructed for serial observation. Volume rendering images were generated for three-dimensional observations.

RESULTS

The mean age of the seven patients was 8.1 years (range, 2–16 years). Four patients were male and three were female. In the present case series, observed CCSFs were all unilateral, and no patient had a family history. Clinical information for all seven patients is shown in Table I.

Medical History

In all cases, the CCSF was discovered at birth and presented with a history of intermittent discharge of watery fluid from the skin opening. The discharge was clear, serous, and increased significantly on eating. One patient (patient 7, Table I) presented with a recent history of recurrent local swelling and pain, which prompted her to seek treatment.

Preauricular appendages were present in all patients on the ipsilateral side and were discovered after birth. Two patients had undergone excision of the preauricular appendages before they were referred to our hospital.

Clinical Symptoms and Signs

All patients showed a punctate skin orifice 1 to 2 cm lateral to the oral commissure (Fig. 1). No palpable mass or tenderness was noted. Local massage or citric acid stimulation could elicit a droplet discharge of clear serous fluid in six patients who showed no infection. In the patient with infection (patient 2, Table I), a 1.0-cm-diameter skin swelling and redness were observed 1.0 cm lateral to the orifice. Intraoral examination showed bilateral normal mucous orifices from Stensen's and Wharton's ducts with normal salivary outflow in all patients. Preauricular appendages were present on the ipsilateral side in five patients. Two patients had undergone surgical excision of the preauricular appendage (patients 2 and 6, Table I). The chin was shifted slightly to the affected side in five patients.

Radiographic Findings

Noncontrast CT and fistulography showed that the skin orifices extended to fistulous tracts and well-defined soft tissue masses in the buccal space. The masses were located anterior to the masseters, superficial to the buccinators, and were filled with contrast medium in fistulography. These masses were presumed to be the ectopic salivary glands (Fig. 2).

Three-dimensional views of the fistulography and parotid sialography showed that the ectopic salivary gland was located inferior to and separate from Stensen's ducts. The Stensen's ducts were intact and, in all cases, showed no communication with the abnormal fistulous ducts (Fig. 3). A normal accessory parotid gland was present in one patient (patient 4, Fig. 3).

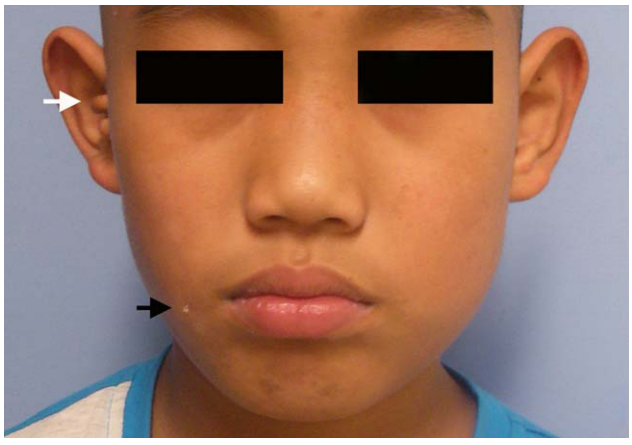

Fig. 1. Clinical image of patient 4. The black arrow indicates the skin orifices of the cheek salivary fistula. Note the saliva secretion elicited with oral stimulation by citric acid. Also note the preauricular appendage (white arrow). Ipsilateral mandibular hypoplasia can be noted and the chin shifts toward the affected side. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

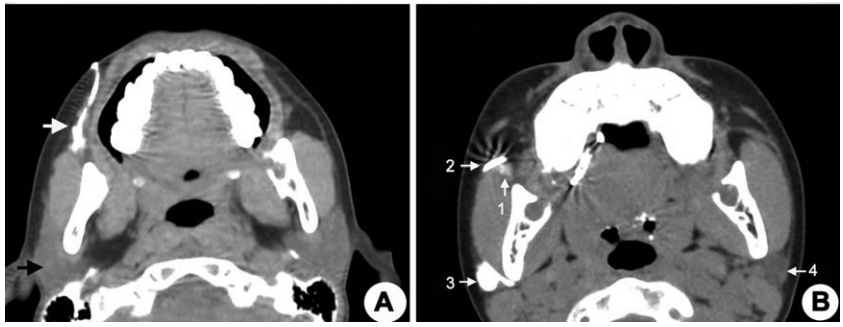

Fig. 2. Cross-sectional computed tomography fistulography (A) and sialography (B) images of patient 4. Axial image of fistulography shows the ectopic accessory salivary gland and its fistulous tract (white arrow) in the buccal space, superficial to the buccinator and anterior to the masseter. Note that the parotid gland and Stensen's duct are not filled with contrast medium during fistulography. (B) Axial image of parotid sialography and fistulography shows that the ectopic gland tissue (white arrow 1) is deep and inferior to and separated from Stensen's duct (white arrow 2). Note the enhanced parotid gland on the ipsilateral side (white arrow 3) and the normal parotid gland on the contralateral side (white arrow 4).

Stensen's duct showed a bifurcation deformity in one case (patient 5).

Three-dimensional views of the mandible showed minor to moderate hypoplasia of the ipsilateral mandibular bodies and ramus in five cases.

Diagnosis and Management

The abnormal soft tissue masses in the buccal space were considered to be redundant ectopic salivary glands on the basis of the saliva-secreting function of the mass associated with eating and citric acid stimulation, and the presence of a normal parotid gland or accessory parotid gland.

Five patients underwent surgical excision of the ectopic salivary gland under general anesthesia. Methylene blue solution was injected through the skin orifice to mark the fistulous tract and ectopic gland. A sliver probe was inserted into the fistulous tract to assist dissection. A buccal mucosal incision was made anteroposteriorly and inferior to the opening of Stensen's duct (Fig. 4A). The ectopic gland and fistulous tract were exposed and dissected to the skin orifice (Fig. 4B). An elliptical incision was used to excise the skin orifice (Fig. 4C). The skin orifice, fistulous tract, and ectopic gland were excised together without injury to Stensen's duct and facial nerve (Fig. 4D). The skin and mucosa incisions were closed.

Histopathologic Findings

Histopathologic examinations of the excised specimens showed that the salivary glands were composed mainly of serous acini, and the fistulous tracts were covered with stratified squamous epithelium.

Follow-up

Follow-up periods for the seven patients varied from 4 months to 6 years. Patients who underwent surgical treatment showed no recurrence of the fistulous discharge or any significant swelling or discomfort during eating. Patients who had not undergone interventional treatment showed no significant progression of the disease.

DISCUSSION

CCSF is very rare, and there are only 10 documented cases in the English literature.[1–9] We have previously reported three cases in two articles.[2,9] This article is the first series study of this syndromic deformity.

Clinical Manifestations

According to previous reports and the present study,[1–9] CCSF occurs with a very distinct set of malformations, and the following common features should be

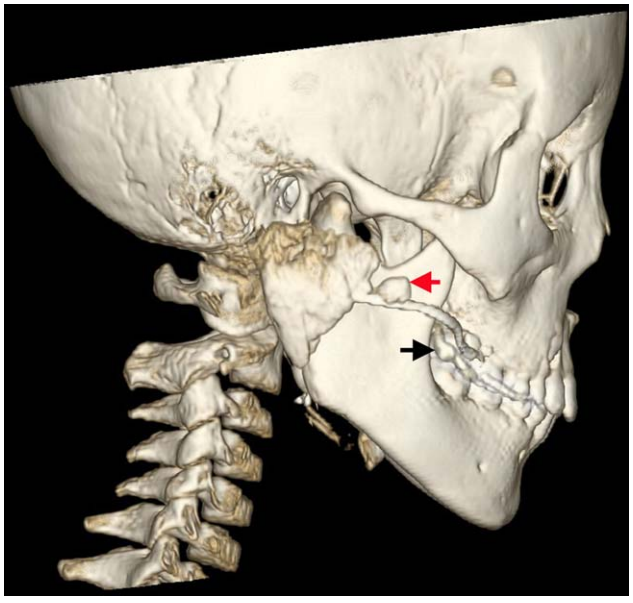

Fig. 3. Three-dimensional computed tomography parotid sialography and fistulography show that the ectopic gland tissue (black arrow) is inferior to and separate from Stensen's duct. Also note the normal accessory parotid gland (red arrow) superior to Stensen's duct. [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

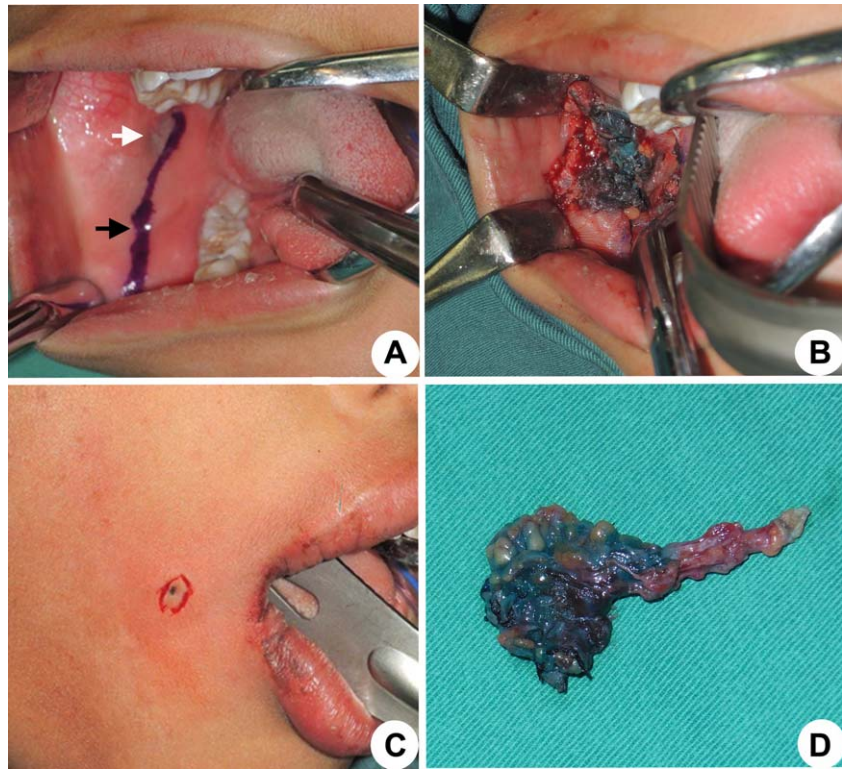

Fig. 4. Surgical procedure. Methylene blue solution is injected through the skin orifice before surgery. A sliver probe is inserted into the fistulous tract. (A) Intraoral buccal mucosa incision (black arrow). Note the mucosal opening of Stensen's duct (white arrow). (B) The ectopic salivary gland, marked by the injected methylene blue, is exposed. (C) An elliptical incision is used to excise the skin orifice. (D) Surgical specimen of ectopic salivary gland with fistulous tract.

noted[2]: 1) congenital cheek saliva-discharging orifice lateral to the oral commissure; 2) presence of ectopic salivary gland connected to the skin orifice through a fistulous tract; 3) the fistulous tract does not communicate with Stensen's duct; 4) the ectopic salivary gland is located in the buccal space anterior to the masseter, inferior to Stensen's duct, and superficial to the buccinators; 5) Stensen's duct can be normal and intact; 6) the presence of ipsilateral preauricular appendage; and 7) ipsilateral hypoplasia of the mandible. Coexistence of the cheek saliva-discharging fistula and preauricular appendage is a prominent clinical feature of this syndrome. The occurrence of CCSF showed no significant gender or side predilection. Only unilateral cases have been documented.

Differential Diagnosis

The skin orifices in CCSF and first branchial fistula both manifest as congenital facial skin fistula but they differ significantly in the following clinical aspects. First, skin orifices of first branchial fistulae usually occur in the parotid and masseter region. CCSF orifices routinely occur close to the oral commissure. Second, first branchial fistulae frequently extend into the parotid gland, to the external auditory canal, and are associated with the facial nerve. The CCSF connects with an ectopic salivary gland inferior to Stensen's duct anterior to the masseter without a direct association with the parotid gland. Third, the first branchial fistula can occur without accompanying malformation.

The term *accessory parotid gland* has been used to describe this salivary gland malformation in previous reports. However, there may be some confusion because the term accessory parotid gland had been used to refer to a common variant of the parotid gland, which is present in about 20% of the population, usually about 6 mm anterior to the main parotid gland.[12] The accessory parotid gland is usually cranial to Stensen's duct with one excretory duct entering Stensen's duct. We suggest using the term *ectopic accessory salivary gland*, because the gland tissue in CCSF is routinely inferior to Stensen's duct and does not enter it.

Etiological Embryogenesis

The occurrence of a preauricular appendage in CCSF is highly indicative of its developmental embryogenesis. The preauricular appendage may occur as an isolated malformation or as part of a generalized syndrome, such as oculo-auriculo-vertebral spectrum.[11] In CCSF, the line connecting the cheek fistula and the preauricular appendage marks the boundary between the union of the mandibular process and maxillary process of the face during embryogenesis. It is reasonably presumed that the CCSF results from abnormal derivation of the first branchial arch apparatus and constitutes a separate variant of the oculo-auriculo-vertebral spectrum.

The auricle develops from the six mesenchymal hillocks around the first branchial cleft. The tragus develops from hillock 1 of the mandibular arch, which is generally close to the angle between the mandibular and maxillary prominences.[13] The auricular appendages are understood to be excess mandibular formations of the hillocks along the edge of the first branchial cleft.[13] At

the time that the auricular appendages appear during the first half of week 6 of gestation, the primordial parotid gland begins to develop as buds from invagination of the inside of the ectoderm of the posterior stomodeum, which proliferate into cords and branches of the gland. This primordial parotid bud is very close to auricle hillock 1. A local mesenchymal stimulus disorder could reasonably give rise to redundant formation of hillock 1 and the salivary primordium. An accessory salivary bud from the outside of the stomodeum could possibly give rise to an additional salivary system separate from the main parotid tissue.

Radiology

CT sialography and fistulography can present accurate three-dimensional views of Stensen's duct, parotid gland, ectopic accessory gland, and its fistulous duct and are essential for appropriate diagnosis and treatment.[2,5] They are also helpful in differential diagnosis with congenital branchial fistula.[14]

Ultrasonography, noncontrast CT or magnetic resonance imaging can help identify ectopic gland tissue in the buccal space, superficial to the buccinators and anterior to the masseter.[1] X-ray sialography and fistulography can also show that the ectopic accessory salivary gland and its fistulous duct are separated from the parotid gland and Stensen's duct,[1,9] but cannot show the three-dimensional view. Magnetic resonance sialography or fistulography using water imaging could possibly generate a three-dimensional image but has not been reported in this situation.

Management

Yamasaki et al.[7] recommend the technique of translocation of the salivary fistulous duct to the oral cavity, during which a tunnel is made through the cheek, and the orifice is implanted into the oral buccal mucosa. However, there is a possibility that the remaining scar in the orifice of the transferred duct might cause an obstruction in the duct. The more conservative treatment of chemocauterization with botulinum toxin has also been reported by Hah et al.[4] A sialocele may occur in the short term after treatment.

In our opinion, surgical excision of the fistula and ectopic salivary gland is effective to control symptoms of salivary discharge in the cheek and prevent recurrence. The following key points are suggested during the surgery: 1) Injection of methylene blue solution through the skin orifice can stain the fistulous tract and ectopic gland effectively, which makes exposure and dissection of the gland and fistula much easier. 2) Inserting a sliver probe into the fistula to indicate the direction of the fistulous tract can ensure complete removal of the fistula. 3) An intraoral approach through the buccal mucosa is useful for dissection of the gland tissue and fistula, during which the incision should be made inferior to the opening of Stensen's duct to prevent injury to the duct. 4) A small elliptical skin incision should be used to excise the skin orifice from an esthetic point of view. These key points are important to prevent surgical injury or postoperative stenosis of Stensen's duct. Other possible complications include hemorrhage, facial nerve injury, and infection. In our series, successful operations and follow-up results were achieved without these complications.

CONCLUSION

A congenital cheek salivary fistula with ipsilateral preauricular appendage constitutes a distinct developmental disorder. Surgical excision can effectively treat congenital cheek salivary fistula.

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