Received: 2 March 2024 | Accepted: 29 August 2024

DOI: 10.1111/odi.15132

ORAL DISEASES Leading in Oral, Maxillofacial, Head & Neck Medicine WILEY

CLINICAL IMAGE

Congenital mandibular appendicular nodule associated with a cervical/submandibular mass

Alfio José Tincani[1] | Gustavo de Souza Vieira[2,3] | Luccas Lavareze[2,3] | João Figueira Scarini[2,3] | Carolina Emerick[2,3] | Pedro Deak de Almeida[1] | Tiago Antônio Baldasso[1] | André Del Negro[1] | André Luís Maion Casarim[1] | Erika Said Abu Egal[3,4] | Albina Altemani[3] | Fernanda Viviane Mariano[3]

[1]Department of Surgery, School of Medical Sciences, State University of Campinas (UNICAMP), Campinas, São Paulo, Brazil

[2]Department of Oral Diagnosis, Piracicaba Dental School, State University of Campinas (UNICAMP), Piracicaba, São Paulo, Brazil

[3]Department of Pathology, School of Medical Sciences, State University of Campinas (UNICAMP), Campinas, São Paulo, Brazil

[4]Biorepository and Molecular Pathology, Huntsman Cancer Institute, University of Utah (UU), Salt Lake City, Utah, USA

Correspondence
Fernanda Viviane Mariano, Department of Pathology - Anatomical Pathology Division, School of Medical Sciences, University of Campinas (UNICAMP), Rua Tessália Vieira de Camargo, 126 – Barão Geraldo, Cidade Universitária, 13083-887, Campinas- SP, Brazil.
Email: fmariano@unicamp.br

1 | CASE REPORT

A seven-months-old female child was referred to the head and neck surgery ambulatory due to the presence of a congenital right mandibular appendicular nodule. The non-consanguineous parents were 23 years-old at birth, after 39 weeks gestation, and no adverse events were reported. On extraoral physical examination, a three centimeters appendicular nodule with a central ostium, secreting a hyaline fluid, was observed in the right mandibular cutaneous region, mobile during breastfeeding (Figure 1a). In addition, an asymptomatic ipsilateral four centimeters cervical/submandibular enlargement was observed at level II (Figure 1b). Intraoral physical examination revealed no changes. Computed tomographic (CT) evaluation demonstrated a well-defined submandibular cystic hypodense image. In addition to these alterations, cardiovascular examinations showed a patent foramen ovale without hemodynamic repercussions.

2 | WHAT IS YOUR DIAGNOSIS?

Based on the patient's history, physical examination, and imaging findings, which one of the following is the most suspicious diagnosis?

A Congenital teratoma.
B Partial diprosopus with developmental cystic formation.
C Cystic hygroma.
D Congenital pyriform sinus fistula.

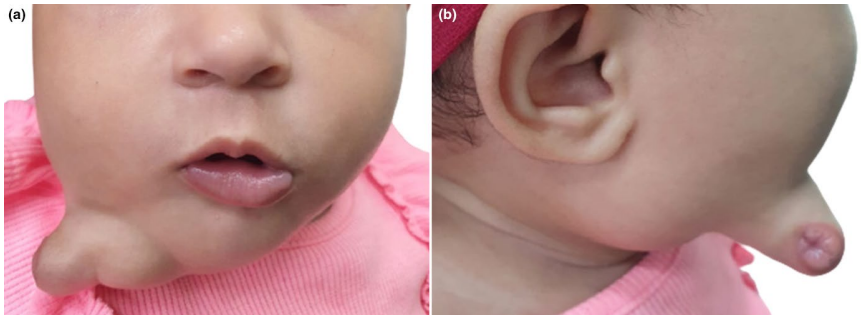

FIGURE 1 Clinical images. (a) Frontal view, showing the appendicular nodule in the mandibular facial region on the right. (b) Lateral view, showing a central ostium to the nodule, associated with an increase in cervical/submandibular volume on the right.

3 | DIAGNOSIS

This patient's condition is partial diprosopus with developmental cystic formation. Diprosopus, also known as craniofacial duplication, is characterized by the duplication of facial structures into a single head, presenting a very low frequency and unknown pathogenesis (Bidondo et al., 2016; Gorlin et al., 2001). Craniofacial duplication can be complete or partial, where a-sided mandibular and mouth duplication are the most common (Morabito et al., 2014). In association with duplicated structures, other maxillofacial conditions may be observed such as cleft lip and palate, and less frequently hamartomas/teratomas and cystic lesions (Costa et al., 2014; Gorlin et al., 2001; Maeda et al., 2013; Mews et al., 2014; Morabito et al., 2014; Paolini et al., 2020).

In our case, through CT scans, a right accessory mandibular segment, showing vascular-nervous canal and tooth germs, was noted. This structure ended toward the cutaneous nodular projection, which was associated with the presence of the central ostium with

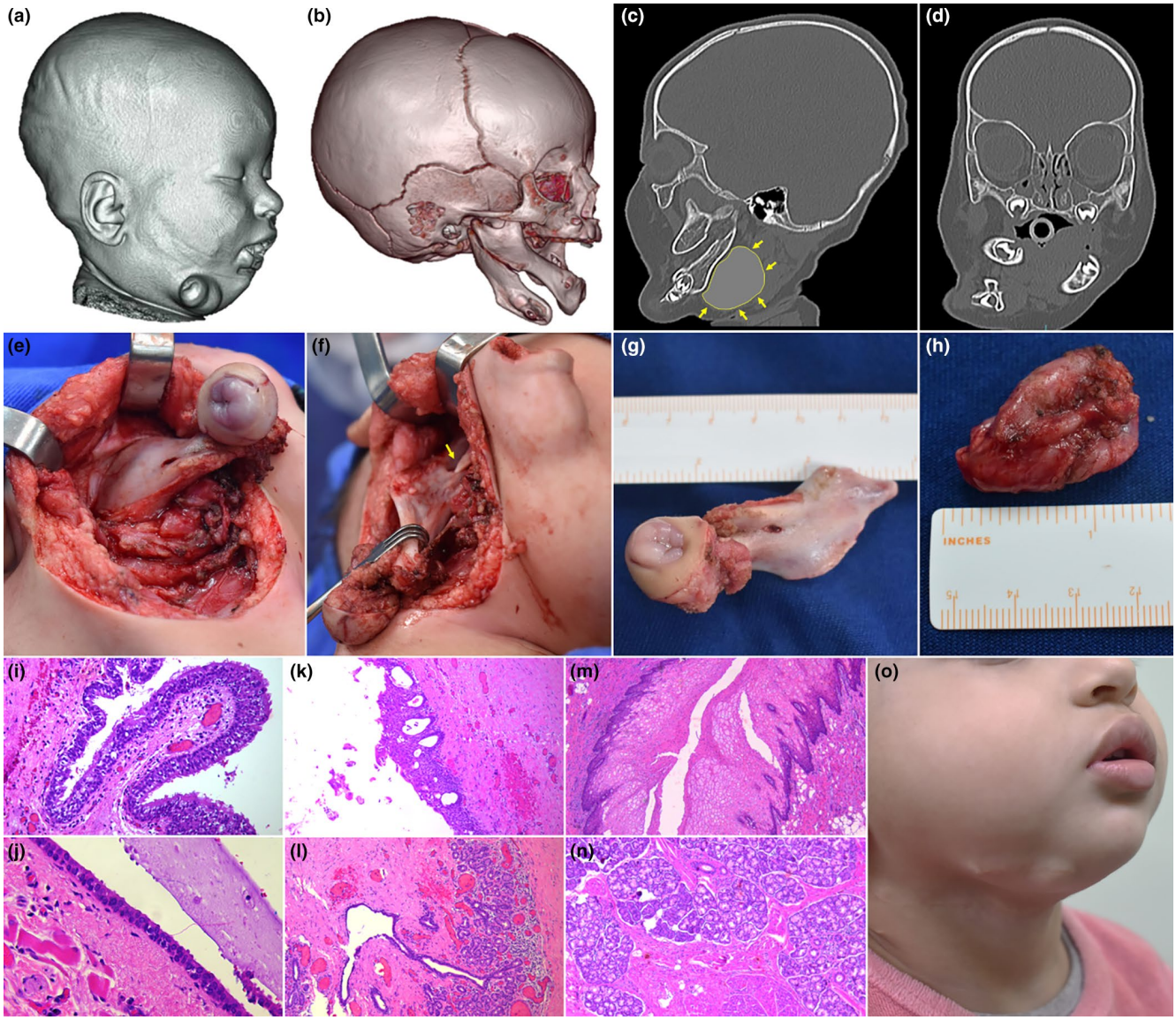

FIGURE 2 Diagnosis and postoperative follow-up. (a, b) 3D CT reconstruction showed the presence of an accessory mandibular segment culminating in the nodular region. (c, d) 2D sections showed the presence of two condyles, mandibular canals in both mandibular segments, normal and anomalous, tooth germs, and an underlying well-demarcated, hypodense, cystic image (arrows). (e, f) Surgical intervention was performed through a submandibular incision to expose the mandibular segment and the cervical/submandibular cyst. Note in detail the penetration of the accessory inferior alveolar nerve into the anomalous mandibular segment (arrow). (g, h) Surgical specimens corresponding to the rudimentary mouth and duplicated mandible (approximately 6.5 cm), and the cystic lesion (approximately 3 cm), respectively. (i–l) Microscopically, the cystic component was lined by pseudostratified cylindrical ciliated, cubic stratified, stratified epithelial patterns with intraepithelial microcystic structures, in addition to the presence of mixed salivary glands in the wall. (m, n) The skin nodule was internally lined with oral mucosa-like tissue, with stratified squamous epithelium with numerous mucous salivary glands in the lamina propria. (o) Postoperative follow-up at 2 years of age showed good healing of the surgical wound.

fluid secretion similar to saliva, and was understood as a rudimentary mouth. Ipsilaterally, in close contact, was a well-defined four centimeters submandibular cystic hypodense image (Figure 2a–d). At 8 months of age, under general anesthesia, the accessory mouth and jaw, as well as the cystic lesion, were excised through an extraoral submandibular approach. Intraoperatively, a bifurcation of the inferior alveolar bundle was evidenced with one segment penetrating each mandibular foramen, normal and accessory (Figure 2e,f). Surgical specimens were sent for anatomopathological examination (Figure 2g,h).

Microscopically, the cystic lesion was lined heterogeneously by cubic, cylindrical ciliated pseudostratified, stratified with pseudoductal structures and stratified squamous epithelia. In the cystic wall, numerous salivary glands were observed. An immunohistochemical study of the cystic lining showed positivity for AE1/AE3, calponin (cytoplasmic), CK7, p63 (basal), and Ki-67 (basal). The rudimentary mouth was internally composed of a stratified squamous epithelium with numerous minor salivary glands in the lamina propria. Given these findings, the submandibular lesion was diagnosed as a developmental cyst associated with maxillofacial anomaly, without further specification. The appendicular nodule was morphologically confirmed as an accessory mouth. The patient is being followed up postoperatively without complications, and the family is receiving genetic counseling for future pregnancies.

Unlike complete diprosopus, the existence of which is mostly incompatible with life, partial forms can be surgically addressed. When gnathic involvement is evidenced, surgical intervention may be postponed until unerupted permanent teeth can be clearly distinguished on imaging examinations, avoiding the removal of dental elements that are part of the normal anatomy. In patients with isolated partial craniofacial duplication without any other associated anomaly, the prognosis is usually favorable after definitive surgery and reconstruction (Hamberis et al., 2020).

4 | OUTCOME

During the postoperative follow-up, there were no complications, and the patient's phonetic and feeding development is as expected. The growth process will be rigorously monitored, and future interventions may be planned for aesthetic and functional purposes, if necessary. The family is receiving genetic counseling for future pregnancies.

AUTHOR CONTRIBUTIONS

Alfio José Tincani: Conceptualization; writing – original draft; methodology; writing – review and editing; supervision. Gustavo de Souza Vieira: Conceptualization; writing – original draft; writing – review and editing; methodology. **Luccas Lavareze:** Writing – original draft; writing – review and editing; methodology. **João Figueira Scarini:** Writing – original draft; writing – review and editing; methodology. **Carolina Emerick:** Writing – original draft; writing – review and editing; methodology. **Pedro Deak de Almeida:** Writing – original draft; writing – review and editing; methodology. **Tiago Antônio Baldasso:** Writing – original draft; writing – review and editing; methodology. **André Del Negro:** Writing – original draft; writing – review and editing; methodology. **André Luís Maion Casarim:** Writing – original draft; writing – review and editing; validation. **Erika Said Abu Egal:** Writing – original draft; writing – review and editing; methodology. **Albina Altemani:** Writing – original draft; conceptualization; methodology; writing – review and editing. **Fernanda Viviane Mariano:** Conceptualization; writing – original draft; writing – review and editing; methodology; supervision.

ACKNOWLEDGMENTS

The São Paulo State Research Foundation (FAPESP, São Paulo, Brazil, grant numbers 19/09419-0; 21/12567-1) and the Coordination of Training of Higher Education Graduate Foundation (CAPES, Brasilia, Brazil, finance code 001).

CONFLICT OF INTEREST STATEMENT

All authors have no conflicts of interest to disclose.

PATIENT CONSENT

The patient reported in this manuscript provided written informed consent for the publication of the case details.

ORCID

Gustavo de Souza Vieira https://orcid.org/0000-0002-3897-6872
Luccas Lavareze https://orcid.org/0000-0003-3464-8879
João Figueira Scarini https://orcid.org/0000-0001-9015-6831
Carolina Emerick https://orcid.org/0000-0001-6904-3242
Erika Said Abu Egal https://orcid.org/0000-0001-9429-6743
Fernanda Viviane Mariano https://orcid.org/0000-0003-3694-5025

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How to cite this article: Tincani, A. J., Vieira, G. S., Lavareze, L., Scarini, J. F., Emerick, C., de Almeida, P. D., Baldasso, T. A., Del Negro, A., Casarim, A. L. M., Egal, E. S. A., Altemani, A., & Mariano, F. V. (2025). Congenital mandibular appendicular nodule associated with a cervical/submandibular mass. *Oral Diseases*, *31*, 341–344. https://doi.org/10.1111/odi.15132