CUTANEOUS SALIVARY GLAND CHORISTOMA IN UNUSUAL SITES

Rajput G.R.[1], Agarwal R.[2], Reddy A.[2]
[1]Department of Dermatology INHS Sanjivani, Kochi, Keralam, India
[2]Department of Dermatology, Venereology and Leprology, Institute of Naval Medicine, INHS Asvini, Colaba, Mumbai, India

Keywords Choristoma, salivary glands, infant.

Abbreviation **HSGT** = heterotopic salivary gland tissue.

Case report. A 5-month-old male infant was brought to our hospital by his mother due to progressively enlarging, erythematous swellings located on the back, left forearm, and left upper arm. The mother first noticed the symptoms when the infant was 2 months old and reported intermittent secretion of a clear, sticky fluid during breastfeeding. Otherwise, the infant was healthy, achieved normal developmental milestones, and presented no congenital anomalies.

Physical examination revealed multiple well-defined, erythematous to flesh-colored, papulonodular lesions measuring 0.5-1.5 cm, distributed across the upper back, left forearm, and left upper arm (Fig. 1, 2). On dermoscopic examination, each lesion exhibited central reddish, structureless areas surrounded by a pale halo and bordered by mild peripheral erythema (Fig. 3). This dermoscopic presentation differed from the description of heterotopic salivary gland tissue (HSGT) provided by Inoue et al., which featured a characteristic red spot surrounded by a pale pink halo with telangiectasias (1).

A punch biopsy revealed well-formed glandular lobules consisting of serous and mucinous acini with ductal elements in the dermis (Fig. 4), demonstrating an architecture identical to normal salivary gland parenchyma observed in typical locations. Immunohistochemical staining for smooth muscle actin (SMA) highlighted the myoepithelial cell layer surrounding the acini and ducts, confirming a salivary gland origin and ruling out adnexal mimics such as eccrine or apocrine tumors (2). Based on clinical and laboratory findings, a diagnosis was made of multifocal salivary gland choristoma of the back, forearm, and upper arm—sites with no known association with the branchial arches.

Fig. 1

Fig. 2

Figs. 1, 2: Cutaneous salivary gland choristoma: erythematous nodular lesions of the back (Fig. 1) and the left upper extremity (Fig. 2).

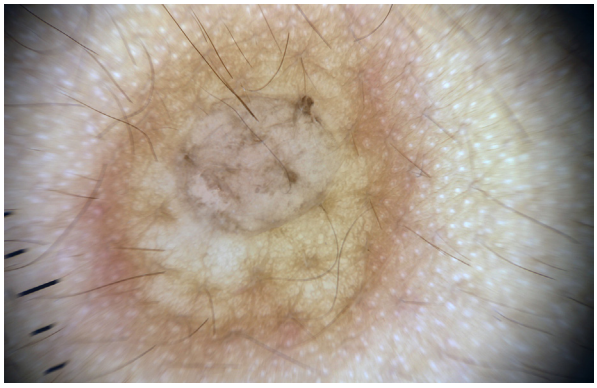

Fig. 3

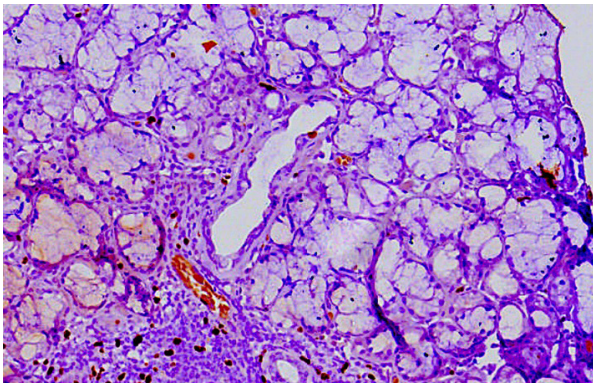

Fig. 4

Fig. 3, 4: Dermoscopic examination (Fig. 3) shows a central structureless area with a peripheral erythematous halo. Histopathological examination (Fig. 4, H&E, 40×) reveals well-organized glandular lobules within the dermis, composed of serous and mucinous acini along with ductal elements.

Discussion. Salivary gland choristoma is an uncommon developmental anomaly characterized by histologically normal salivary tissue situated in an aberrant location (3). Intermittent secretion of a clear, sticky fluid during breastfeeding is considered a classic diagnostic sign of cutaneous heterotopic salivary gland tissue (HSGT). This occurs due to the parasympathetic salivary reflex, which stimulates both ectopic and native salivary glands during feeding (4). Cutaneous manifestations typically present as draining sinuses or papules located at the angle of the mandible, and are believed to arise from the entrapment of glandular tissue during branchial arch fusion (4, 5). Rarely, authors have reported the presence of aberrant salivary glands in sites other than the head and neck, including the middle ear, pituitary gland, mediastinum, gastrointestinal tract, and vulva (6, 7).

Our literature search identified a limited number of published cases of cutaneous HSGT. The largest series, described by Haemel et al. (4), included 11 patients with cervical draining sinuses; all lesions were confined to the neck, with a predilection for the right side. Other reports have similarly localized cutaneous HSGT to the preauricular, periparotid, or mandibular angle regions (5, 8). To the best of our knowledge, only three previous cases describe a cutaneous salivary gland choristoma outside the head and neck: Shin CB et al. (9) reported a chest wall HSGT in a 6-year-old girl; Aby et al. (10) reported a chest wall choristoma in a newborn; and Marwah et al. described an aberrant salivary gland mass in the vulva (11).

Cervical HSGT is explained by entrapment during the fusion of the first and second branchial arches; however, this mechanism cannot account for locations distant from the branchial apparatus. A hypothesized explanation could be the aberrant caudal migration of neural crest cells, which normally contribute to the myoepithelial and stromal cells of the salivary glands (6, 12). Another possibility is that multipotent cutaneous adnexal stem cells differentiated toward a salivary gland lineage; however, such lesions would typically exhibit a mixed cellular pattern on histology. The well-organized lobular architecture in our case argues in favor of a true choristoma rather than a metaplastic change. While chest wall lesions have been attributed to the anomalous migration of branchial arch tissue along the milk line, the lesions on our patient's back and upper extremities cannot be explained solely by branchial arch embryogenesis.

Recognizing HSGT is important for two reasons. First, correct identification allows it to be classified as a benign condition, thereby avoiding further invasive interventions. Second, malignant transformation into mucoepidermoid carcinoma, acinic cell carcinoma, and adenocarcinoma has been documented in 6 out of 24 cases (25%) of HSGT in one series (13), although nearly all occurred in adults

and in non-cutaneous sites. Therefore, long-term follow-up is advisable, particularly for atypical presentations. The feeding-related secretion observed in our patient and described by other authors (1, 14) serves as a valuable clinical clue.

In **conclusion**, we report the first case of multifocal cutaneous salivary gland choristoma involving areas beyond the cervicofacial region. The diagnostic triad consisting of erythematous papulonodules, feeding-related secretion, and SMA-positive glandular tissue on histopathology is highly reliable. Clinicians should consider this diagnosis when evaluating draining papules in infants, regardless of their anatomical location.

Conflicts of interest

The authors declare that they have no conflicts of interest.

Address to:
Dr. Gopalsing Rameshsing Rajput, MD (Derm, Ven and Lep)
Associate Professor
Department of Dermatology, Venereology and Leprology,
INHS Sanjivani
Kochi, Keralam, India

References

1) Inoue S, Maeda T, NagataY, et al. Dermoscopic features of heterotopic salivary gland tissue in the neck: report of a case. *Case Rep Dermatol*. 2024;16(1):17-20. doi: 10.1159/000535740.
2) Zarbo RJ, Regezi JA, Batsakis JG. S-100 protein in salivary gland tumors: an immunohistochemical study of 129 cases. *Head Neck Surg*. 1986;8(4):268-75. doi: 10.1002/hed.2890080406.
3) Willis RA. The borderland of embryology and pathology. 2nd ed., Butterworths, London 1963.
4) Haemel A, Gnepp DR, Carlsten J,, Robinson-Bostom L. Heterotopic salivary gland tissue in the neck. *J Am Acad Dermatol*. 2008;58(2):251-6. doi: 10.1016/j.jaad.2007.11.009
5) Ferlito A, Bertino G, Rinaldo A, et al. A review of heterotopia and associated salivary gland neoplasms of the head and neck. *J Laryngol Otol*. 1999;113(4):299-303. doi: 10.1017/s0022215100143841.
6) Pesavento G, Ferlito A. Benign mixed tumor of heterotopic salivary gland tissue in the upper neck. Report of a case with a review of the literature on heterotopic salivary gland tissue. *J Laryngol Otol*. 1976;90(6):577-84. doi: 10.1017/s0022215100082475.
7) Lassaletta-Atienza L, López-Ríos F, Martín G, et al. Salivary gland heterotopia in the lower neck: a report of five cases. *Int J Pediatr Otorhinolaryngol*. 1998;43(2):153-61. doi: 10.1016/s0165-5876(97)00173-0.
8) Ha SL, Shin JE, Yoon TH. Salivary gland choristoma of the middle ear: a case report. *Am J Otolaryngol*. 2000;21(2):127-30. doi: 10.1016/s0196-0709(00)85011-5.
9) Shin CE, Kim SS, Chwals WJ. Salivary gland choristoma of the anterior chest wall. *J Pediatr Surg*. 2000;35(10):1506-7.
10) Aby Jl, Patel M, Sundram U, Benjamin LT. Salivary gland choristoma (heterotopic salivary gland tissue) of the anterior chest wall of a newborn. *Pediatr Dermatol*. 2014;31(1):e36-7. doi: 10.1111/pde.12159.
11) Marwah S, Berman ML. Ectopic salivary gland in the vulva (choristoma): report of a case and review of the literature. *Obstet Gynecol*. 1980;56(3):389-91. PMID: 7422183
12) Emmerson E, Knox SM. Salivary gland stem cells: A review of development, regeneration and cancer. *Genesis*. 2018;56(5):e23211. doi: 10.1002/dvg.23211.
13) Daniel E, McGuirt WF Sr. Neck masses secondary to heterotopic salivary gland tissue: a 25-year experience. *Am J Otolaryngol*. 2005;26(2):96-100. doi: 10.1016/j.amjoto.2004.08.009.
14) Jain S, Aggarwal A, Deshmukh P, et al. Heterotopic salivary gland presenting as a discharging sinus in the base of the neck. Clin Pract. 2011;1(4):e131. doi: 10.4081/cp.2011.e131.