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Ectopic parotid: case report

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SUMMARY: **Ectopic parotid: case report.**

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A recent case led the authors to re-examine the clinical characteristics of the cervical ectopia of the major salivary glands. These glands develop in the embryo between the sixth and seventh week, starting with the formation of endodermal invaginations of the branchial section of the floor of the primitive mouth. These cell cords, initially solid, proliferate in the underlying mesenchyme, starting from the opening of the future excretory duct, and subsequently branch and canalize.

During embryogenesis, the endodermal invaginations become closely interconnected with the adjacent lymphatic tissue. It is thus possible for lymphoid tissue to migrate into the parotid or the other major salivary glands, or conversely, for salivary tissue to become included in the cervical lymph nodes. Very rarely, ectopic salivary gland tissue can also be found in other unusual locations, including the neck region, as a result of a developmental abnormality of the branchial apparatus. The base of the neck is the most common location, while ectopia of the mid third of the neck is quite rare.

The authors discuss the clinical details and diagnostic procedure leading to preoperative diagnosis. This congenital anomaly can, albeit rarely, degenerate into cancer, and surgical excision is thus imperative.

RIASSUNTO: **Ectopia parotidea. Considerazioni su un caso clinico.**

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Gli Autori prendono lo spunto da un caso clinico, giunto di recente alla loro osservazione, per discutere le caratteristiche cliniche dell'ectopia cervicale delle ghiandole salivari maggiori. Tali ghiandole si sviluppano tra la sesta e la settima settimana di vita embrionale, in seguito alla formazione di invaginazioni endodermiche del pavimento della sezione branchiale della bocca primitiva. Questi cordoni cellulari, dapprima solidi, proliferano nel mesenchima sottostante a partire dall'orifizio di sbocco della futura sezione escretrice e, successivamente, si ramificano e si canalizzano.

Durante l'embriogenesi si instaurano intime relazioni tra le predette invaginazioni endodermiche ed il tessuto linfatico adiacente con conseguente possibile migrazione di tessuto linfoide all'interno della parotide e delle altre ghiandole salivari maggiori o, viceversa, di tessuto salivare che risulta incluso nei linfonodi cervicali. Molto raramente tessuto ghiandolare salivare ectopico può essere rinvenuto in altre sedi inusuali, tra cui le altre regioni del collo, come risultato di una anomalia di sviluppo collegata all'apparato branchiale. L'ectopia localizzata alla base del collo è sicuramente la più comune, mentre sono alquanto rari i casi rinvenibili a carico del terzo medio. Premesse le ipotesi patogenetiche, gli Autori definiscono le caratteristiche cliniche e l'iter diagnostico che possono portare ad una diagnosi pre-operatoria. Concludono affermando che detta anomalia congenita non consente di escludere la possibilità di una degenerazione neoplastica, seppur estremamente rara, e sostengono che l'escissione chirurgica sia l'unico atto che nel contempo completi sia l'iter diagnostico che il percorso terapeutico.

KEY WORDS: Parotid gland - Ectopia - Surgery.
Ghiandola parotide - Ectopia - Chirurgia.

Introduction

The major salivary glands develop in the embryo between the sixth and seven week, following the formation of endodermal invaginations in the floor of the branchial section of the primitive mouth. These cell cords, initially solid, proliferate in the underlying mesenchyme, starting

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from the opening of the future excretory duct, and subsequently branch and canalize.

During embryogenesis, the salivary glands become closely interconnected with the adjacent lymphatic tissue. It is thus possible for lymphoid tissue to migrate into the parotid or the other major salivary glands, or conversely, for salivary tissue to become included in the cervical lymph nodes, which are therefore the most common location for ectopic salivary gland tissue (1).

Ectopic salivary gland tissue may also very rarely be found in other unusual locations, such as the middle ear (2), soft tissues of the neck and mouth (3), pineal gland (remnant of Rathke's pouch) (4), thyroglossal duct, thyroid gland (5), parathyroid gland (6), tonsils (7), cerebellopontine angle (8), and even areas far from the head and neck such as the rectum (9), stomach, and vulva (10).

The recent observation of an ectopic parotid gland led us to re-examine the embryogenesis behind the onset of this congenital abnormality, as well as its clinical characteristics.

Case report

This case concerned a 57-year-old man, who had nothing of particular interest in his family or medical history. His past history included just a total thyroidectomy for a benign disease, carried out around nine years previously.

About six months before coming to our attention, the patient had suffered an episode of fairly mild pain lasting just a few hours during which he noticed a swelling in the left anterolateral area of the middle third of the neck, descending to the inner edge of the sternocleidomastoid muscle. Clinical examination revealed an approximately oval, taut, elastic swelling, with maximum diameter 2.5 cm and indistinct margins, behind the inner edge of the sternocleidomastoid muscle. The swelling was quite well anchored to the underlying tissue.

Ultrasound examination revealed only some localized reactive lymphadenopathies on the left. CT scan found asymmetry of the oropharyngeal and laryngeal lumen due to the presence of a swelling in the left posterolateral area of the base of the tongue and the ipsilateral hypopharynx, and hyperdense nodules in the former area of the left thyroid lobe. These findings suggested the presence of residual thyroid tissue. On ENT examination the oropharynx and pharynx appeared normal, with no evident signs of mucosal inflammation or lesions. The examination confirmed the presence of a moderate swelling of the left posterolateral wall of the hypopharynx, covered with normal-looking mucosa. Finally, MRI revealed both the sequelae of the previous thyroidectomy and a bulk on the left with intermediate signal intensity in T1, and slight hyperintensity in T2. After contrast administration the swelling showed homogeneous enhancement, tentatively compatible with residual thyroid tissue, without excluding other diagnostic possibilities (Figs. 1,2). It was therefore decided to proceed to surgical exploration.

A 3-cm incision was made parallel to the medial edge of the left sternocleidomastoid muscle. After spreading apart the layers of the strap muscles, a multilobular neoplasm was discovered descending behind the lateral margin of the trachea, appearing to be continuous with a parathyroid gland. A short cord, looking very like an excretory duct, originated from this growth. The neoplasm and duct were

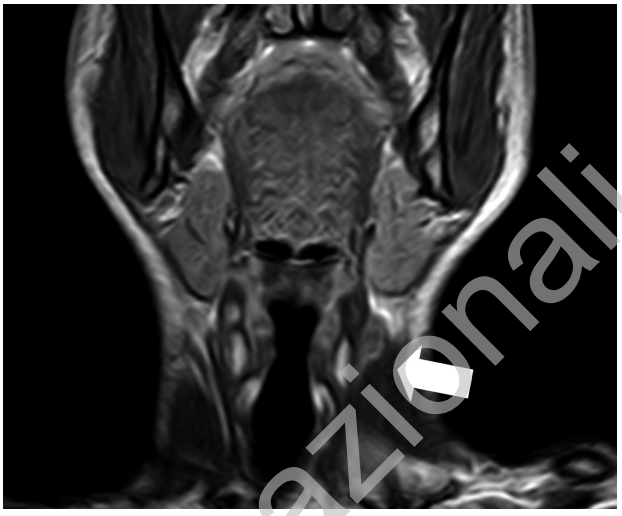

Fig. 1 - MRI. Nodular formation in the left posterolateral area of the neck.

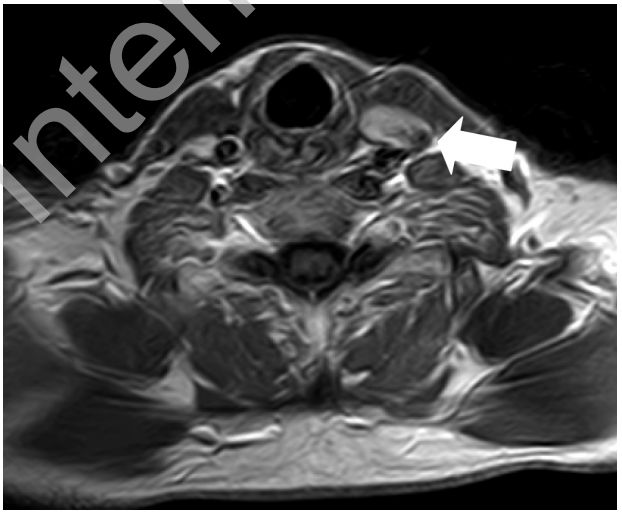

Fig. 2 - MRI. After contrast enhancement, homogeneous impregnation of the formation.

carefully isolated and removed. After assuring hemostasis, the incision was closed in layers.

Macroscopic examination of the excised tissue confirmed that it was a multilobular neoplasm of 5 x 3.5 x 1.5 cm, of gland-like appearance, which on cutting revealed a moderately sized excretory duct (Fig. 3). Histological examination found it to be composed of compound, acinar glands with serous secretions, with moderate fatty involution and slight focal lymphocytic infiltration. The excretory ducts appeared normal. The morphology of the acinar cells, which contained abundant zymogen granules, seemed completely preserved (Figs. 4, 5). These histological findings were thus structurally indistinguishable from normal parotid parenchyma.

Discussion

The finding of an ectopic parotid gland adjacent to the parathyroid glands is a particularly rare event, with

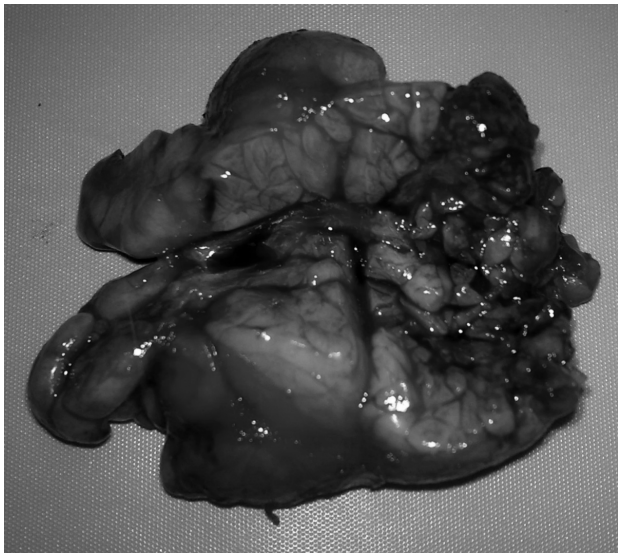

Fig. 3 - Multilobular formation with glandular appearance containing an excretory duct.

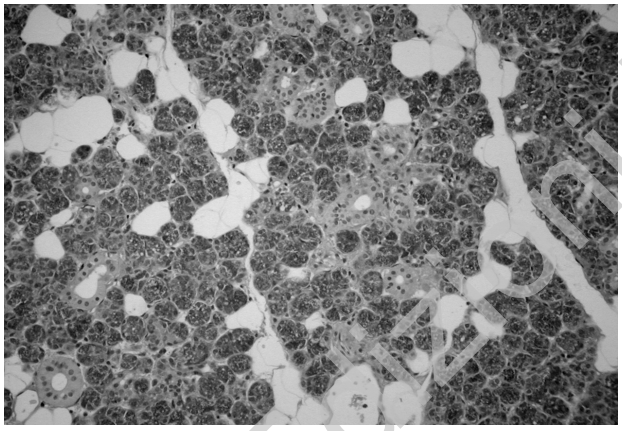

Fig. 4 - Acinar gland consisting of serous secretion, with intact, undilated excretory ducts.

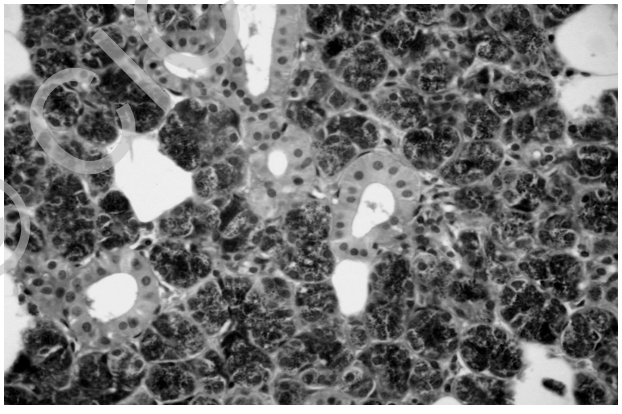

Fig. 5 - At higher magnification, preserved acinar cells containing abundant zymogen granules.

few reports in the literature. In a review of 759 histological preparations of parathyroid glands, Edwards and Bhuia (11) described two cases of salivary gland foci found inside the parathyroids, both associated with cystic formations. Carney (6) described five cases of peri-parathyroid salivary ectopia associated with cysts; of these, four were found in patients operated for hyperparathyroidism and the fifth in a patient operated for a thyroid nodule. Finally, a review of histological preparations of all parathyroid glands excised from 258 patients at the Mayo Clinic in one year found two cystic formations with associated salivary ectopia.

The etiopathogenesis of ectopic cervical salivary glands is still debated. The unusual location and association with other malformations presenting as sinuses, fistulas, cysts or even cartilage reinforce the theory of a connection with developmental abnormalities of the branchial apparatus. Youngs and Schofield (12) proposed the most plausible theory, suggesting that the ectopia is due to an unusual persistence and differentiation of the endodermal remnants of the precervical sinus of His. After the second week of embryonic life, the second branchial arch hypertrophies, incorporating the third and fourth arches. The second branchial cleft thus deepens and the endodermal surface of arches II-V flattens out, while the inner surface forms a temporary cavity called the cervical sinus.

Ectopic salivary glands in the neck most frequently emerge from the anterior triangle along the medial edge of the sternocleidomastoid muscle (13). The base of the neck is the most common location, while literary reports of glandular ectopia in the middle third of the neck (14), as in our case, are rare. A high neck location (with the exception of ectopic tissue included in the lymph nodes) is almost always associated with salivary gland tumors (pleomorphic adenoma, mucoepidermoid carcinoma). Ectopic salivary glands present as swellings of the subcutaneous tissue, and consist of sinuses that generally open externally, secreting an odorless liquid similar to saliva, especially during meals. They are often found during childhood or adolescence, and in rare cases may be bilateral or associated with congenital abnormalities of the branchial apparatus (15).

In branchial malformations it can happen that one arch becomes prematurely fused to the next, enclosing an ectodermal fold covered with squamous epithelium and producing a dermoid cyst. Alternatively, the inclusion may only involve the endodermal groove, covered with ciliated epithelium, giving rise to a mucoid cyst. In other cases, the second arch does not fuse completely to the wall and the cervical sinus persists over a greater or lesser path. This leads to a branchial fistula which is usually complete, opening onto the surface of the neck and passing behind the sternocleidomastoid to the hyoid bone, where it descends between the two carotids, opening in-

ternally in the pharynx. The sinuses almost always open externally in the laterocervical area.

From a clinical perspective, this gives rise to the need for differential diagnosis between an ectopic salivary gland in the neck and other abnormalities originating from the branchial arches, clefts and pouches, especially lateral subhyoid cysts and fistulas of the neck originating from the cervical sinus or the second (or exceptionally the third) pharyngeal pouch. These conditions do in fact have some common features:

- same origin, as both branchial lesions and ectopic salivary glands are the result of a congenital malformation;
- no functional disorders, slow growth, generally small size (diameter 1-5 cm);
- possible location of the external opening along the medial edge of the sternocleidomastoid muscle;
- possible presence of an intermittent secretion, and no pain, except during episodes of bacterial superinfection.

The differential features are as follows:

- no history of infection for ectopic salivary glands, whereas lateral branchial cysts and fistulas of the neck have a strong tendency to suppuration;
- increased salivary secretion, especially during meals or while chewing, is a constant feature of salivary ectopia.

The differential diagnosis should also consider other possible disorders such as cystic hygroma, benign or malignant lymphadenopathy, lymphomas, hemangiomas, neurofibromas, etc. Naturally, metastasis of a salivary gland neoplasm or a tumor originating in the ectopic salivary tissue must not be excluded (16-20).

Instrumental diagnosis is often not fully definitive, as in our case. However, it is generally conclusive in the case of branchial cysts, which on ultrasound present as clearly delimited anechogenic masses. CT is also important for the diagnosis of branchial cysts, which typically appear as clearly circumscribed and homogeneously hypodense. MRI above all enables a more accurate diagnosis, revealing the depth of the lesion. Where a fistula has been observed, injection of a radiopaque dye helps differential diagnosis by establishing its path and the underlying structures:

- cystic dilation and possible communication with the pharynx in the case of branchial sinuses, fistulas and cysts of the neck;
- treelike structure in the case of ectopic salivary glands.

Conclusion

The development of the salivary glands begins with proliferation of the ectodermal (parotid) or endodermal (sublingual and submandibular gland) epithelial tissue. While the parathyroids migrate during embryonic development, the tissues that give rise to the salivary glands do not, and are limited to the area around the jaw.

In our case, given the absence of any branchial malformation (sinus, cyst or similar) associated with the lesion, the presence of glandular tissue in this area could be explained by the theory that during embryo development there is dislocation of the aberrant glandular tissue originating from the pharyngeal pouches, along with a part or bud of the final organ, and its differentiation into salivary tissue at the site of the ectopia. This is supported by the fact that the parathyroid glands originate from buds that lose their connection with the epithelium of the pharyngeal pouches before migrating to their final location. In this area, the epithelium of the dorsal expansion of the third and fourth pouch differentiates into the lower and upper parathyroids.

Histologically, this is almost always normal salivary gland tissue, although the remote possibility of a tumor originating from the ectopic tissue should always be borne in mind. For this reason, we believe that these lesions should be treated by complete excision under elective surgery. This is especially important in cases where the diagnostic procedure has not removed all doubts as to the possible nature of the neoplasm, given that surgical removal and histological examination alone are often conclusive, as in our case.

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