CASE STUDY

Mammary Analogue Secretory Carcinoma of Salivary Glands: Report of Clinical Case

Carcinoma análogo del secretor mamario de glándulas salivales: a propósito de un caso clínico

Carlota Rovira, a, Xavier León, a Celina P. Vásquez, b Montserrat López a

a Servicio de Otorrinolaringología, Hospital de Sant Pau, Barcelona, Spain
b Servicio de Anatomía Patológica, Hospital de Sant Pau, Barcelona, Spain

Received 14 March 2017; accepted 29 June 2017

A 29-year-old woman with no disease history of interest consulted with a painless right infra-auricular mass of one month’s onset with no other clinical symptoms. FNA revealed moderately atypical glandular cells, and occasional squamous cell groups with no accompanying stroma. The report was positive for malignant cells.

MRI reported the presence of a nodule, 14 mm × 16 mm in the posterior area of the right parotid tail, of discretely hyperintense contour on T2 with a centre of lower signal intensity, well-defined limits hypointense on T1, suggestive of a benign parotid tumour (Fig. 1).

A parotidectomy I–II was performed together with partial resection of the sternocleidomastoid muscle, as the tumour was adhering to it. The result of the perioperative biopsy was compatible with mucoepidermoid parotid carcinoma, and therefore a radical neck dissection was performed of right area II.

The deferred pathological study showed a low-grade parotid carcinoma with morphological features of a mammary analogue secretory carcinoma (MASC), 1.2 cm in size.

Macroscopically, the tumour had well-defined borders with focal infiltration of the glandular parenchyma. Histologically polylobulated, microcystic and tubular architecture was observed, eosinophil secretions and an area of central hyalinisation, with occasional mitosis figures. There were no areas of necrosis or vascular or perineural invasion. Immunohistological staining was positive for mammaglobin, GCDFP-15, 5100 vimentin, PAS and cytokeratin 7. The FISH study showed translocation t(12;15)(p13;q25). The resection limit of the sternocleidomastoid muscle was negative, and the right area II dissection harvested 12 lymph nodes with no evidence of neoplasm (Fig. 2).

With a diagnosis of low-grade MASC treated with appropriate resection, the Committee of Head and Neck Tumours did not consider adjuvant treatment necessary.

Discussion

MASC of salivary glands is a new tumour entity described for the first time in 2010 by Skálová et al. Its most characteristic feature is the presence of translocation t(12;15)(p13;q25), that results in the fusion of the ETV6-NTRK3 gene that codifies a chimeric thyrosin-quinase protein with epithelial, mesenchymal and blood cell transforming potential.1 This tumour shares histological and genetic features with mammary secretory carcinoma, a type of slow-growing, low-grade, mammary tumour that occurs in young women.2


* Corresponding author.
E-mail address: carlotarovirar@yahoo.com (C. Rovira).

2017 Sociedad Española de Otorrinolaringología y Cirugía de Cabeza y Cuello. Published by Elsevier España, S.L.U. All rights reserved.
The most common site is the parotid gland (70%), followed by the submandibular gland and the oral cavity. The incidence is unknown, principally due to classification errors prior to its description in 2010, although some studies put it at around 4% of the malignant tumours of the salivary glands. It is slightly more predominant in males than females (53%).

Onset is usually in the 4th decade of life, with a wide range of ages (15-70 years).

This type of tumour usually presents as a painless, slow-growing nodule with indolent clinical course.

On contrast CT, the lesion appears well-circumscribed with moderate contrast uptake. On MRI, the lesion is usually hypointense on T1 in relation to the muscle, and hypointense in T2 in relation to the parotid tissue.

Diagnosis is usually confirmed by histological, immunohistochemical and molecular study of the surgical specimen. The usual macroscopic structure is a well-circumscribed mass, subdivided by fibrous septa. Macroscopically the mass has tubular, papillary or solid microcystic-type architecture. The cells usually show acinar or mucoid differentiation with vacuolated, eosinophilic cytoplasm, colloid-like intraluminal and/or intracellular secretions positive for PAS staining.

The cell nucleus is mild to moderately typical, with low mitotic activity. Immunohistochemical study shows high positivity of S100, and specific positivity for mammaglobin, a protein derived from uteroglobin. It is also positive for broad spectrum cytokeratins (AE1/AE3), low molecular weight (CK7, CK8 and CK19), CDFP-15, MUC1, protein 3, α-amylase, DOG-1,P63, STAT5a, and vimentin.

Definitive diagnosis is based on molecular study showing the presence of translocation t(12;15)(p13;q25), by FISH or by detection of ETV6-NTRK3 fusion transcript with RT-PCR.

The differential diagnosis of low-grade cases must principally include acinar cell carcinoma, especially the variant with few cytoplasmatic granules, mucoepidermoid carcinoma and cystoadenocarcinoma. Differential diagnosis of high-grade cases, must include ductal cell carcinoma.

The treatment of choice for MASC is surgical resection with appropriate limits. Because it has more of a tendency to metastasise to the lymph nodes than other low-grade carcinomas, neck dissection should be considered, although there is insufficient data to support systematic neck dissection in clinical N0 patients. Adjuvant radiotherapy is reserved for cases where the margins are positive, or near to the tumour (<5 mm), incomplete resections, perineural invasion, T3-T4 tumours, and cases with positive
adenopathies. Anti-TRK thyrosin-kinase inhibitors (resulting from ETV6-NTRK3 fusion) could be considered for cases that are not candidates for surgery, since they have shown powerful activity against this thyrosin-kinase in vitro, entrectinib especially.

The prognosis is usually favourable, although in high-grade cases, the clinical course is much more aggressive, and the prognosis poorer.

Conflict of Interests

The authors have no conflict of interests to declare.

References


