

Mammary Analogue Secretory Carcinoma of the Parotid Gland: Report of a case of a newly characterised salivary gland tumour

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Abstract

Mammary Analogue Secretory Carcinoma is a new entity in salivary gland tumours that has only been recently described. It has overlapping histopathological features with acinic cell carcinoma and low grade variants of cystadenocarcinoma. It has a distinct chromosomal translocation, t(12;15)(p13;q25), resulting in a fusion gene ETV6 NTRK3. The clinical course of this disease entity is uncertain at present. We present a case of MASC of the parotid gland occurring in a 34 year old man, and discuss the difficulties in diagnosis and subsequent treatment of this tumour.

Case report

A 30 year old previously healthy man presented with a painless left cheek mass that had been slowly growing over the past six months. On physical examination there was a 2cm firm, non tender left sided parotid mass that was not fixed to surrounding structures. There were no palpable enlarged cervical lymph nodes. The facial nerve had normal function. The rest of the clinical examination was unremarkable.

A fine needle aspiration cytology was performed on the left cheek lump and this was reported as a "cystic epithelial neoplasm with atypia".

Radiologic findings

A computerized tomographic (CT) scan revealed a 1.6 x 2.0 cm heterogenous enhancing mass lesion with hypodense cystic areas seen within the superficial lobe of the left parotid gland (Fig.1) The radiologic findings were suggestive of a Warthin's tumour or a pleomorphic adenoma.

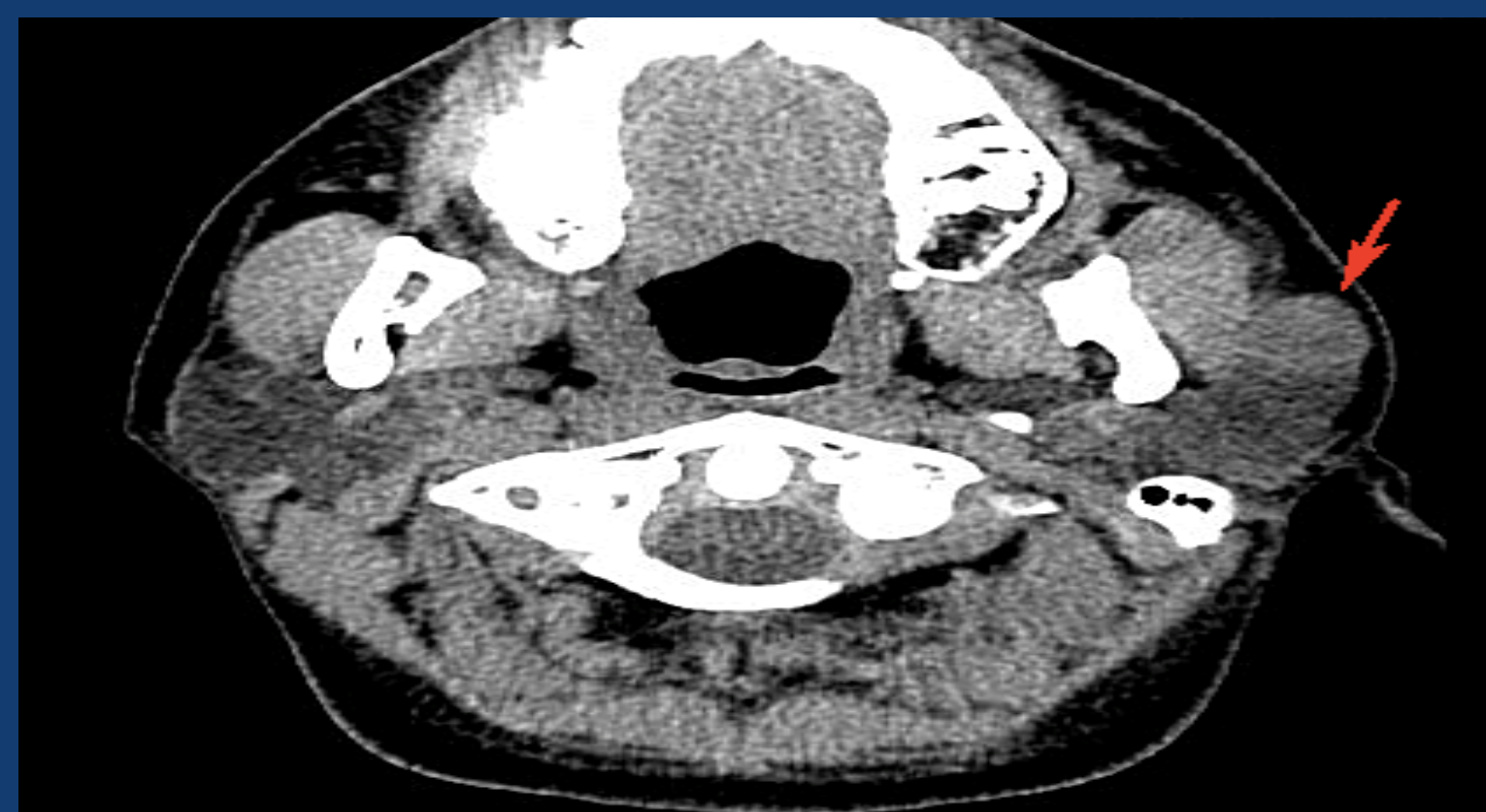


Fig.1

Operative findings

A left superficial parotidectomy with preservation of the facial nerve was performed after a 2 month delay due to the patient. During these 2 months, the tumour had been growing significantly and the size was 5 cm at the day of surgery, but remained painless.

Intraoperatively, surgical dissection was difficult because of the large size of the tumour and needle decompression was attempted to facilitate resection and identification of tissue planes. Yellowish fluid and blood clots were aspirated from the tumour. The facial nerve was identified and this was found to be wrapping around the tumour. Surgical dissection was difficult as tumour was in close proximity to the facial nerve and small amount of tumour tissue was left behind around the buccal and zygomatic branches of the facial nerve in a bid to preserve as much facial nerve function as possible.

Postoperative Magnetic Resonance Imaging scans showed no residual tumour.

Results

Gross pathological features:

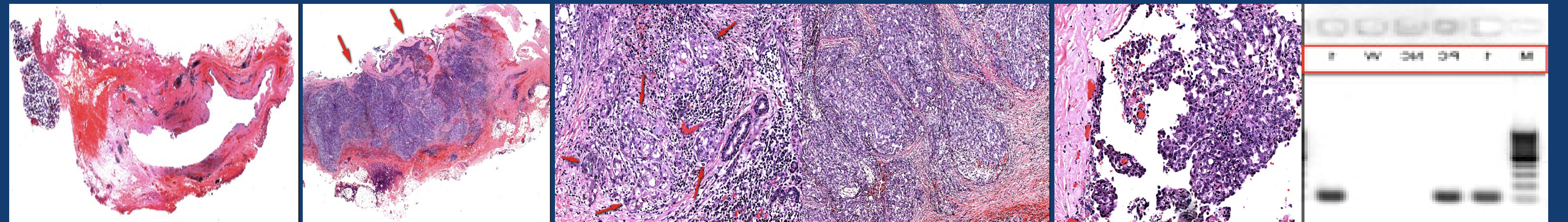
The resected specimen was fragmented and the tumour displayed both cystic and solid components.

Histology, Cytology and Immunohistochemistry:

The microscopic examination confirmed a salivary gland neoplasm with cystic and solid areas (Fig. 2A,B). In the non-cystic areas clearly invasive growth was identified (Fig. 2C). Apart from the macrocystic areas, several other different growth patterns; solid and pseudociriform, microcystic as well as papillary areas, were identified (Fig. 2D,E). The neoplastic cells had round to oval nuclei containing pale pink granular to vacuolated cytoplasm. Secretory material was present within the microcystic spaces and was positive for PAS (periodic acid-Schiff). There was no necrosis and the mitotic activity was low (<1 / 10 high power fields). Tumour margins were positive. The tumour cells displayed strong immunoreactivity for S100 protein in the immunohistochemical study.

Molecular genetic study:

A rearrangement of ETV6 was documented in the FISH study and an ETV6-NTRK3 gene fusion was documented on RT-PCR (Fig. 2F)



Figs 2A-F

Discussion

Mammary analogue secretory carcinoma (MASC) is a new neoplastic entity in salivary gland pathology which was first described by Skalova et al in a series comprising 16 salivary cases. A few small series and case reports have since been published. The majority these cases has occurred in the parotid, although it has also been found to affect the submandibular and minor salivary glands. There is a gender predilection for males, majority of whom are in their 40s-50s.

Diagnostic challenges arise from several aspects. The cytological characteristics of MASC are unknown by most cytopathologists. The invasive component of low-grade salivary gland carcinomas frequently also does not give rise to radiologically identifiable invasion.

Histologically, from a differential diagnostic point of view, there is overlap between MASC and mainly zymogen poor acinic cell carcinoma (ACC) and low-grade variants of cystadenocarcinoma (CACA). However, only MASC shows ETV6 rearrangements and the presence of the ETV6-NTRK fusion transcript. Although histologically, MASC shows similarities to these low grade salivary gland neoplasms, some cases of MASC have pursued an aggressive course and postoperatively, may recur locally and give rise to both regional lymph node and distant metastasis. Our patient was recently diagnosed, precluding any meaningful follow-up.

To date, there is limited data on the clinical behaviour of MASC and the diagnosis and management of such patients is not straightforward. Increased awareness of this recently characterised malignant salivary gland neoplasm is likely to improve the quality of preoperative investigations and management of patients with this tumour.