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Case report

Unusual presentation of parotid gland adenoid cystic carcinoma : A case presentation and literature review ☆,☆☆

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ABSTRACT

Adenoid cystic carcinoma (ACC) represents approximately 10% of all epithelial salivary neoplasms and most commonly involves the parotid gland.

We report CT and MRI finding of a 38-year-old young man presented to our ENT department with 02 years history of an external auditory canal stenosis. Physical examination revealed bilateral parotid gland swelling with a complete stenosis of the left external auditory canal.

Temporal bone contrast enhanced CT-SCAN revealed is an isodense enhancing mass measuring 4 cm involving posterior and inferior external auditory canal wall, and invading the superficial lobe of the homolateral parotid gland. No bone erosion was noted. MRI investigation has shown a tumor process highly suspicious of malignancy centered on the left EAC involving the superficial lobe of the homolateral parotid gland. Adenoid cystic carcinoma (ACC) of the parotid gland was the final diagnosis, after surgical biopsy and histopathological examination. The Pre-operative check-up demonstrated multiple round shape lung lesions suggestive of multiple metastases. Considering the metastatic stage of the tumor, a collegial decision to adopt a palliative treatment approach based on chemotherapy was taken by the multidisciplinary oncology board.

☆ All authors agreed for publication

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Introduction

Cystic adenoid carcinomas or cylindromas were first described by Robin and Laboulbene in 1852 [1]. The cystic adenoid carcinomas are rare and usually develop in salivary glands. They represent the main histological type of accessory salivary gland malignancies and account for approximately 10%-20% of all salivary gland tumors [2]. Adenoid cystic carcinomas represent about 1% of ENT cancers, but may more rarely occur in all sites with secretory glands (breast, cervix, colon, prostate, etc.) [3]. Despite a mild histological appearance and satisfactory therapeutic results in the short term [4], the management of ENT cylindromas remains delicate due to their insidious growth, their neurological tropism and their metastatic potential. These factors together delay diagnosis, which is often made when they are locally advanced. Surgery is difficult and late recurrences are frequent [4].

Case presentation

A 38-year young man without no significant history or drug use, presented to our ENT department following the discovery of an external auditory canal stenosis evolving over the last 2 years.

Physical examination revealed a mild bilateral parotid homogeneous gland swelling with a complete soft tissue stenosis of the left external auditory canal (EAC). No ear discharge or facial nerve paralysis were noticed and no cervical lymphadenopathy were noticed.

A temporal bone CT scan with MRI investigations were performed to evaluate the origin and the extension of the left EAC process. A contrast enhanced temporal bone CT scan showed a homogeneous enhancement of a soft tissue process that fills completely the left external auditory canal, and

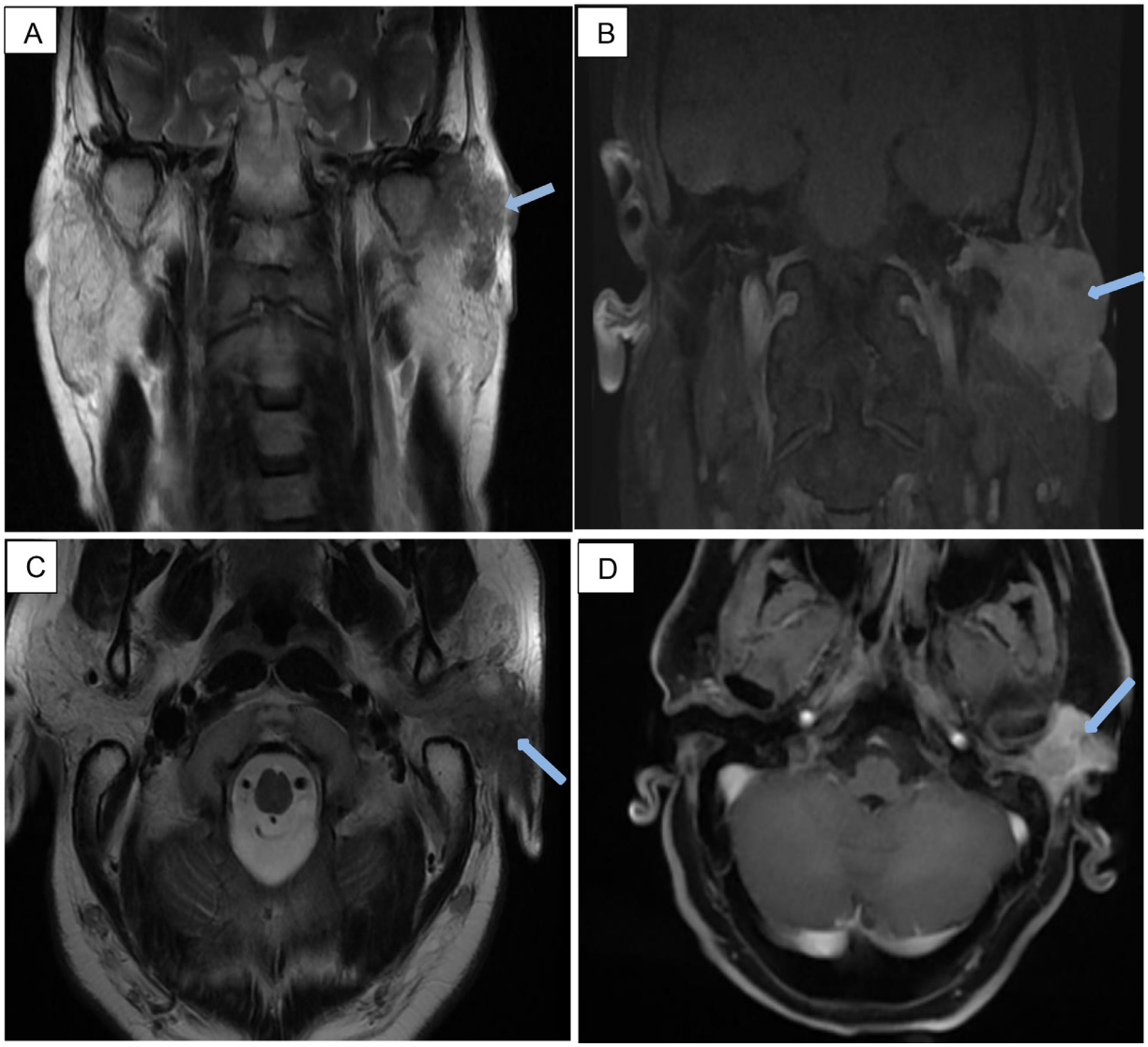

Fig. 1 – A well-defined lobulated soft tissue mass invading the auricle, the superficial lobe of the left parotid gland and the posterior inferior wall of the EAC. It appears as an intermediate signal intensity on T2WI (A, B), with heterogeneous enhancement following IV contrast (C, D). There is no perineural invasion, nor loco regional lymphadenopathy. The irregular margins and infiltrative appearance suggest a higher-grade malignancy.

involve also the homolateral parotid gland superficial lobe, with no extension to the middle ear, no associated bone destruction, and no adjacent cerebral parenchymal abnormality (Fig. 1)

On MRI this mass appeared as a tumor of highly suspicious malignancy arising from the left ACE and invading the superficial lobe of the homolateral parotid gland. The tumor seemed mainly centered on the EAC (Fig. 2).

The preoperative assessment revealed multiple secondary pulmonary locations, without other secondary lesions (Fig. 1 C).

A surgical biopsy of the left EAC and the parotid gland tumor was performed under general anesthesia. Specimen pathology examination confirmed the diagnosis of adenoid cystic carcinoma (Figs. 3, 4). At the MDM (multidisciplinary oncology meeting) the decision was to start a chemotherapy based on cisplatin and vinorelbine since the patient had a general metastatic disease.

Discussion

Cervicofacial adenoid cystic carcinomas (ACC) are relatively rare malignant epithelial tumors (less than 1% of cervicofacial malignancies). They develop mainly in accessory and main salivary glands, lacrimal glands and mucous glands of the

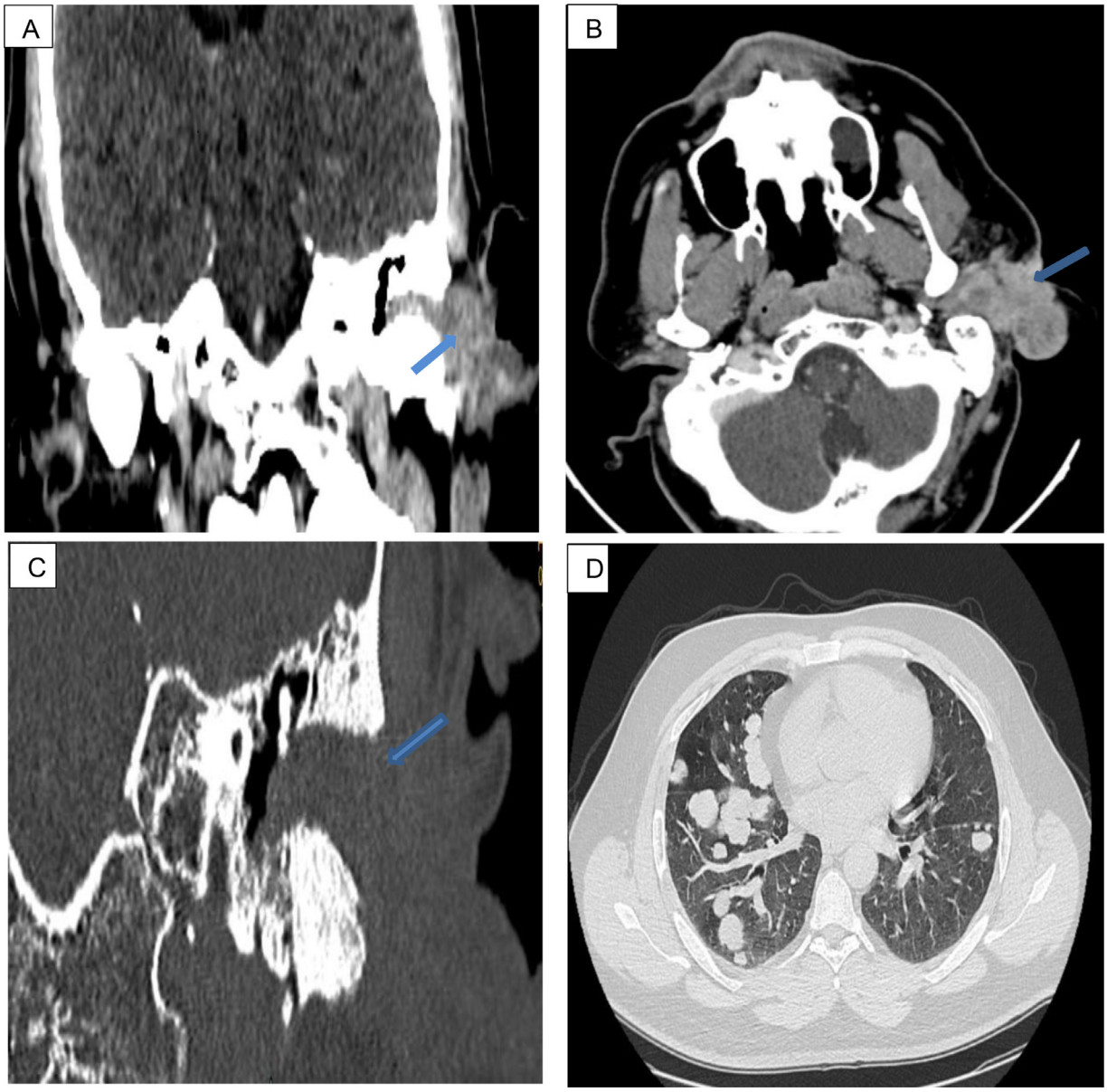

Fig. 2 – Tissue process heterogeusly enhanced, which appears to originate from the posterior inferior wall of the EAC, invading the auricle and the superficial lobe of the homolateral parotid gland (A; B), with no involvement of the middle ear, non-associated bone lysis (C), and no adjacent cerebral parenchymal abnormality.
The preoperative assessment revealed multiple secondary pulmonary locations, without other secondary lesions (D).

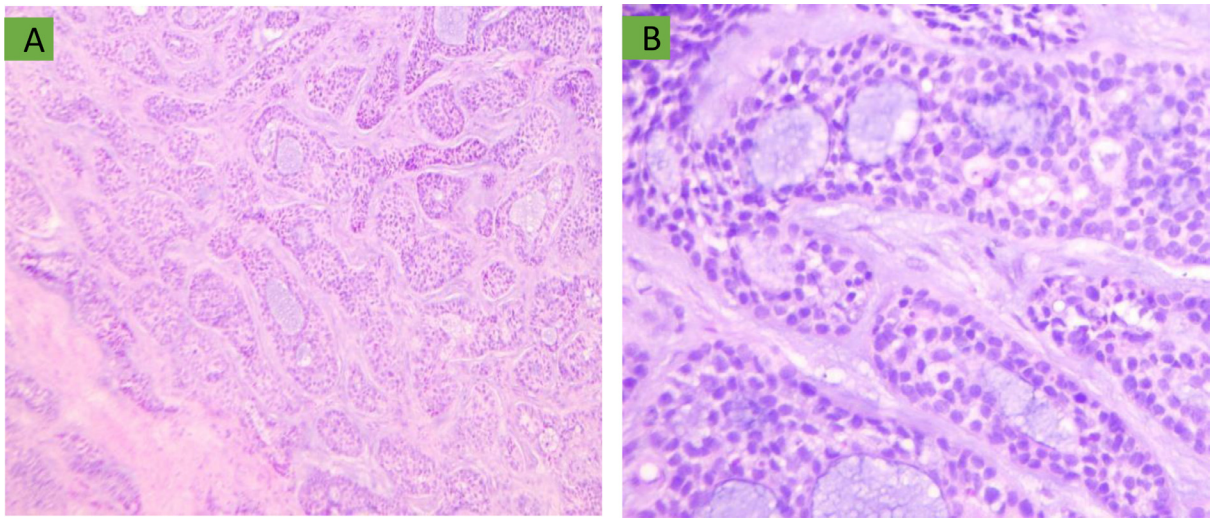

Fig. 3 – Carrot biopsy showing tumour proliferation consisting of cribriform clusters and irregular trabeculae, centred by hyaline material
HESx100 (A); HESx200 (B).

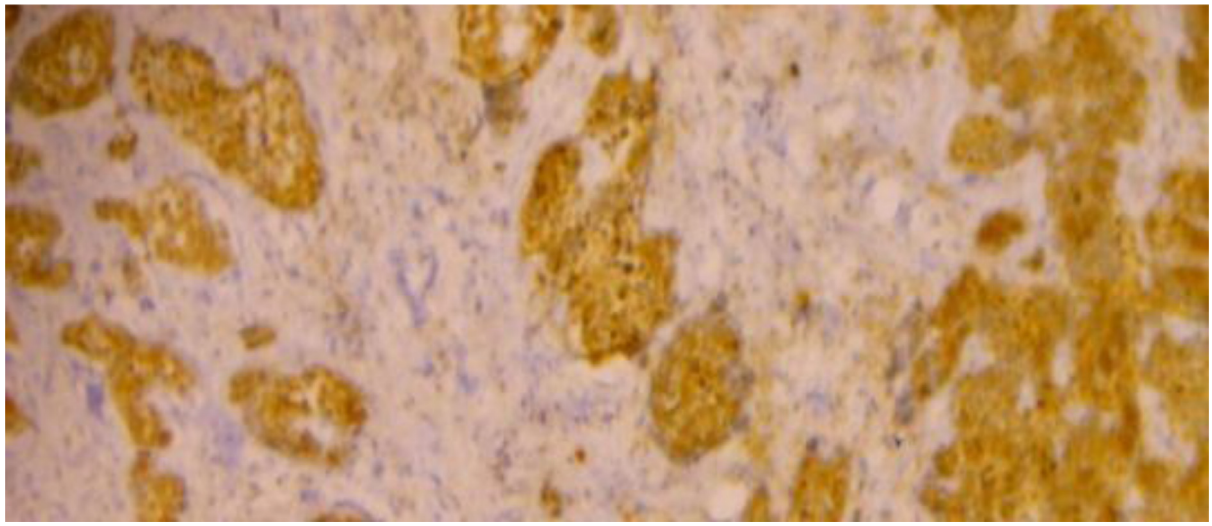

Fig. 4 – Immunohistochemical studies showing that most of the tumor cells are immunopositive for CK.

nasal cavity and paranasal sinuses. These tumors account for 5% of all tumors of the main salivary glands. The parotid gland is the most frequently affected [3,4] ACC is one of the most common and well-limited malignancies of the salivary glands.

The term "adenoid cystic carcinoma" was described by Ewing in 1954 [5]. This tumor was named "cylindroma" earlier by Billroth in 1859 because the epithelial and connective elements formed a system of intertwined cylinders. The term "basalioma" was first noticed by Krompecher in 1908, [6] who considered this type of tumourtumor to be analogous in nature to the growth of basal cells in the skin. [6] It is a relentless tumor that is prone to local recurrence and possibly distant metastases. [5] When the tumor arises from the parotid gland, it presents as a swelling or a solid mass, sometimes associated with a dull pain and/or a paralysis of a cranial nerve, in particular the facial nerve [1]. The main features of this type of neoplasm are peri-neural invasion, which occurs in approximately 22% to 46% of cases, and multiple local recurrences .In our patient the involvement of the EAC could be related to tumoral spread through it sensory nerves such as the greater auricular nerve of the superficial cervical plexus or due to the involvement of some branches of the facial nerve particularly the auricular posterior branche which is directed to the superior and posterior auricular muscles however, the patient presented neither left auricular anesthesia or paresthesia nor facial paralysis. Regional lymph node involvement is considered rare. However, distant metastases have an incidence of 40% [7], and can affect different sites, most commonly the lungs, bones and liver [2].

Histopathologically, ACC has 3 forms: cribriform, tubular, and solid. The cribriform glandular pattern is the most common and best recognized appearance. It is marked by islands of basaloid epithelial cells that contains multiple cylindrical areas [3] Commonly, ACC is composed of cribriform and tubular patterns [8]. All variants can show a prominent perineural invasion, and the tumor can follow the course of a nerve for a long distance; the neoplasm can also show intraneural invasion, considered an independent negative prognostic factor [9,10].

Preoperative imaging is considered as the key modality to approach the diagnosis, to establish the extent of theses tumors, and generally used in all post treatment follow up procedures. Ultrasound examination shows no specific signs to distinguish ACC from other neck neoplasms. However, Ultrasound-guided needle aspiration cytology can be helpful to distinguish malignant from benign lesions, but the accuracy of this procedure is defendant on the operator's experience, with a sensitivity of 88%- to 93% and a specificity of 75%- to 99% [11]. The main goal of diagnostic imaging is to assess the anatomical extension of the tumor, which is crucial for an accurate surgical planning. Obviously, CT-scan can better delimit bone invasion, while MRI is preferable to approach the nature of the lesion, and to evaluate the locoregional extension across the deep planes, and the infiltration of the cervical lymph nodes and bone marrow [12]. Imaging studies of the skull base is mandatory to investigate the intracranial extension through the retrograde peri-neuronal pathway, is as important as its caudal extension to the cervico-thoracic passage [12].

The MRI imaging techniques includes conventional morphologic T1 weighted and T2 weighted sequences, as well as Diffusion Weighted Imaging (DWI) sequences. ACC can appear as a defined mass or an ill-defined mass with diffuse infiltration of its surrounding structures; it's usually homogeneously enhanced after contrast media- injection, although heterogeneous enhancement due to necrosis may be noticed [12]. The solid and more cellular histological subtype of ACC has lower signal on T2-weighted MRI imaging [13]. Irregular margins, adjacent tissue infiltration and hypo intensity in T2-weighted sequences are characteristic of salivary gland carcinoma, respectively with decreasing predictive value [13]. Apparent Diffusion Coefficient (ADC) allows distinguishing between ACC and pleomorphic adenoma, but has a low predictive value of malignancy [14]. Dynamic Perfusion-Weighted (PWI) sequences increase MRI sensitivity for carcinomas, but not specificity; ACC often shows a rapid wash-in form plateau, which is also typical in pleomorphic adenoma, but with much lower ADC values [15]. A recent study by Singh et al. [16] described all the imaging features of peri-neural tumor spread: enlargement and/or erosion of foramen, nerve enlargement and/or enhancement, obliteration of the peri-neural fat tissue layer, including pterygopalatine fossa (PPF), enlargement and convexity of the lateral cavernous sinus wall, soft-tissue replacement of cerebrospinal fluid Meckel's cave, muscular denervation. particularly, muscular denervation can be considered a secondary sign of nerve damage: firstly, in acute and subacute stages, oedema appears, while the chronic appearance is characterized by fatty replacement of muscle tissue and by muscular atrophy [17]. However, the possibility that many other conditions, such as infection, inflammation, trauma, vascular lesion, and hematoma can mimic a neoplasm of the head and neck region must be considered [19]. sensitivity of MRI is superior to CT scan (95% -to 100%) in detecting ACC's perineural spread along the skull base, but the sensitivity, when mapping the extent of disease, decreases to 63% [18]. CT-scan is complementary to MRI in the study of local bone changes of the skull-base foramina [19].

Moreover, MRI represents the reference standard for post-therapy follow-up, while whole-body CT scan and 18F-FDG PET-CT scan can be used to detect distant metastases, particularly in initial staging and in post-treatment monitoring [18,20]. however, the cornerstone of treatment is surgery, while radiotherapy has been considered for advanced stages and as adjuvant in the presence of positive microscopic margins. A total conservative or a radical parotidectomy is advocated for tumors occurring in parotid, though the main intent is to obtain a tumor-free area of at least 1 cm [21]. Because metastasis to regional lymph nodes is uncommon, neck dissection is typically not indicated [22]. Some authors suggest that advanced and non-resectable tumors may be treated only with radiotherapy.

Conclusion

ACC is a rare malignant tumor of the parotid gland. The main goal of treatment in patients with CCA is local control, normal functionality and prevention of distant metastases. Metastases can occur very late, and therefore long-term follow-up and a high index of suspicion are necessary for early diagnosis to allow a more favorable prognosis and better quality of life. However, this case is particularly interesting because of the initial presentation of the disease. In fact, the 2 years history and the absence of a clinical well defined Parotid tumor has lured either the radiology specialist end clinician to consider this tumor as primarily arising from the EAC. Unfortunately, given the fact that the patient has a general metastatic stage all curative therapies could not be proposed.

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