Giant adenoid cystic carcinoma of the nasal cavity

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INTRODUCTION

We report an unusual case of giant adenoid cystic carcinoma developed in the nasal cavity. Based on a brief literature review, we discuss the clinical, imaging, and histological features of this tumor, as well as available treatments and prognostic factors.

CASE-REPORT

A 64-year-old patient from a rural part of Morocco presented to the ENT emergency department in Rabat with a large mass located in the nasal area, which had developed about 1 year earlier and grown at a rapid pace. The mass measured 14 cm along its greatest axis and was implanted on the nose and right paranasal area. It was multilobate, firm, smooth, and painless (Figure 1). She reported insidious development of bilateral nasal obstruction over the last 18 months. Other manifestations included anosmia and a purulent, blood-tinged, and fetid nasal discharge. The patient was in good general health. Anterior rhinoscopy showed a budding tumor filling the right nasal cavity and displacing the septum into the left nasal cavity, closing it entirely. Ophthalmological examination showed epiphora in the right eye with a cataract and decreased vision, as well as lateral displacement of the right eyeball with telecanthus.

Computed tomography (CT) of the face and sinonasal cavities disclosed a large mass of tissue density with contrast enhancement. The mass was located in the right nasoethmoidal cavities and extended to the right maxillary sinus and right orbit, displacing the right eyeball laterally without invading it. The tumor was in contact with the skull base but did not extend inside the skull. A major mass effect on the nasal septum was noted (Figure 2). Examination of an endonasal biopsy specimen established the diagnosis of adenoid cystic carcinoma with a cribriform pattern (Figure 3). Investigations found no evidence of distant metastases.

En-bloc tumor resection was performed via the right paralateronasal approach. This left a defect in the right lateral aspect of the nose requiring reconstruc-

Figure 1: Exteriorized mass

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tion with a paramedian forehead flap. The immediate postoperative course was uneventful. The flap pedicle was divided after 2 weeks. Histological examination of the operative specimen confirmed the diagnosis of adenoid cystic carcinoma. The margins were deeply invaded. External-beam radiation therapy was delivered to the midface and sinonasal cavities, in a dose of 60 Grays with a conventional fractionation schedule. No macroscopic recurrence was found at follow-up 6 months later.

Figure 2: Computed tomography of the sinonasal area, axial section with contrast medium injection: ***huge heterogeneous high-density protruding mass filling the right nasal cavity and extending to the right maxillary sinus.***

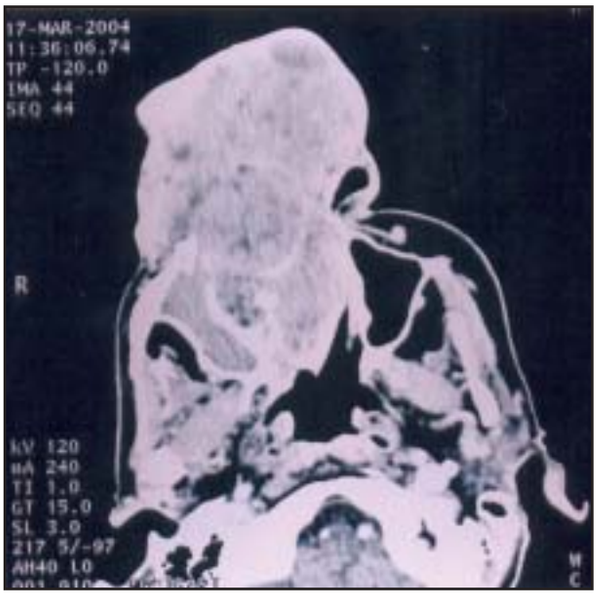
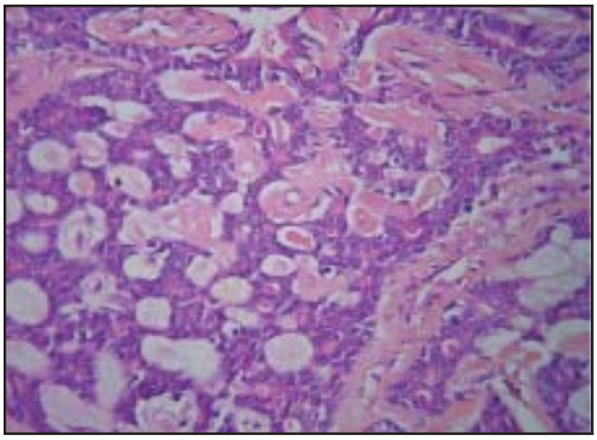


Figure 3: Adenoid cystic carcinoma: ***cribriform lobules composed of somewhat irregular cells scattered in a fibrohyaline stroma. HE, x250.***



DISCUSSION

Adenoid cystic carcinomas contribute 10% of all salivary gland tumors [1]. They usually arise in accessory salivary glands, most notably those located in the oral cavity. Few cases of development in the sinonasal cavities have been reported. Adenoid cystic carcinomas usually grow slowly and are advanced when diagnosed. The fast rate of growth in our patient is unusual, but we are not aware of earlier reported describing similarly exteriorized tumors.

Although the course may be favorable initially, many patients subsequently experience local recurrences and remote metastases, with selective lung involvement. As a result, the 10-year survival rate is less than 10% [2]. The local recurrence rate is very high in patients with sinonasal involvement, among whom 60% experience clinically patent recurrences within 2 years after treatment [3]. This reflects the difficulty in achieving tumor-free margins at the skull base: the tumors are often advanced, the region is anatomically complex, endocranial tumor spread along the cranial nerves is common, and the proximity of critical nervous and vascular structures limits the extent of resection. The main prognostic factors include tumor stage, histological grade (with a better prognosis for cribriform and tubular tumors than for solid tumors), whether perineural spread is present, and whether the resection margins are positive [2,4].

En bloc resection is essential but difficult to achieve near the skull base [2,5]. Postoperative radiation therapy improves the long-term prognosis in patients with large tumors, most notably when microscopic tumor remnants are present after surgery. It has been suggested that postoperative radiation therapy should be given routinely to patients with sinonasal adenoid cystic carcinomas [1-2]. Fast neutron therapy may produce better outcomes than conventional radiation therapy in patients with macroscopic remnants [5]. Full-dose preoperative radiation therapy has been used in patients with T4N0M0 tumors to prevent cancer cell dissemination during surgery [2]. Given the potential for progression, long-term follow-up with clinical evaluations and imaging studies is mandatory.

CONCLUSION

Adenoid cystic carcinomas of the sinonasal cavities are aggressive tumors associated with high rates of local recurrent and distant metastases, regardless of

the treatment modalities used. Combining radiation therapy and surgery increases the chances of disease control.

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