A case of highly aggressive adenoid cystic carcinoma of the external auditory canal

Carcinoma adenoido cistico del canale uditivo esterno: descrizione di un caso

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Key words

External auditory canal • Glandular tumours • Carcinoma • Surgical therapy

Parole chiave

Condotto uditivo esterno • Tumori ghiandolari • Carcinoma • Trattamento chirurgico

Summary

Tumours of the external auditory canal are extremely rare and only 20% of these are of glandular origin. The most frequent histotype is adenoid cystic carcinoma. The rarity of external auditory canal glandular tumours explains the lack of large series reported in the literature and the corresponding large number of case reports from different Authors. Adenoid cystic carcinoma, presenting in the external auditory canal, exhibits the same characteristics as those affecting the major salivary glands, this tumour has an aggressive behaviour characterized by local invasivity and with a metastatic risk of approximately 30%. A rare case of adenoid cystic carcinoma of the external ear is reported. The patient, a 75-year-old male, had right intermittent otorrhea for 6 years. On examination, a vegetating, ulcerated formation which easily bled was found protruding from the right external auditory meatus. Clinical, radiological and pathological features of the tumour are described. A subtotal petrosectomy combined with homolateral elective lymph node neck dissection was performed. Parotid gland, condyle of the mandible and VII cranial nerve were spared since these were free from disease.

Riassunto

I tumori del canale uditivo esterno sono estremamente rari e, di questi, soltanto il 20% hanno origine ghiandolare. L'istotipo che più frequentemente è stato riscontrato è il carcinoma adenoidocistico. La rarità dei tumori del canale uditivo esterno spiega la mancanza di un'ampia casistica in letteratura ed il corrispondente numero elevato di casi singoli riportati da vari Autori. Il carcinoma adenoidocistico del canale uditivo esterno ha le stesse caratteristiche di quello che colpisce le ghiandole salivari maggiori, ha un comportamento caratterizzato da invasività locale ed un rischio metastatico di circa il 30%. È decritto un raro caso di carcinoma adenoidocistico del canale uditivo esterno. Il paziente, maschio di 75 anni, lamentava otorrea intermittente da circa 6 anni. All'esame obiettivo si valutava la presenza di una massa sanguinante a livello del meato acustico destro. Sono decritti i risultati degli esami clinici e radiologici eseguiti. La terapia chirurgica è stata caratterizzata da una petrosectomia con dissezione dei linfonodi latero-cervicali. La parotide, il condilo mandibolare ed il nervo faciale sono stati risparmiati perché indenni dalla patologia.

Introduction

Tumours of the external auditory canal (EAC) are extremely rare and only 20% of these are of glandular origin [1].

The most frequent histotype is adenoid cystic carcinoma (ACC) [2 3]. Indeed, during the period 1928-1980, of 395,000 neoplasms examined at the Presbiterian Hospital in New York, only 25 appeared to be glandular tumours of the EAC and 10 of these were ACC [4]. The rarity of EAC glandular tumours explains the lack of large series reported in the literature and the corresponding large number of case reports from various Authors [5-13]. ACC presenting in the EAC exhibits the same characteristics as those affecting the major salivary glands and may have a tubular, cribriform or solid pattern, the latter related to the worst prognosis [4 14].

This tumour has an aggressive behaviour characterized by local invasivity. As for the ACC presenting in the salivary glands, the metastatic risk is approximately 30% while cutaneous locations have a lower metastatic risk (8%) [13].

It is interesting to note that the origin of this tumour, following the hypothesis of Perzin et al. [4], has been related to heterotopic salivary remnants rather than major salivary glands extension [15].

Glandular neoplasms of the EAC show a male preponderance (M/F ratio = 2:1), the major incidence being in the white race and with a wide age range (18-87 years, mean 42) [2 15] being more aggressive in younger ages. The symptoms are hypoacusis, otorrhea and pain. The latter symptom is almost exclusively related to neural invasion in ACC. Due to local aggressivity, the resection margins are of great importance [16 17], indeed, several Authors suggest obtaining

routinely frozen sections to evaluate local and perineural invasion [4 18]. The 5-year survival rate is high [4] but the local recurrence rate is equally high.

Case report

The patient, a 75-year-old male, had right intermittent otorrhea for 6 years. On examination, a vegetating, ulcerated and easily bleeding formation was found protruding from the right external auditory meatus while the auricular pinna and the retroauricular sulcus were infiltrated. Examination of biopsy tissue revealed an infiltrating epithelial tumour. High resolution computed tomography (CT) scan with contrast medium of the temporal bone showed a huge formation involving the mastoid, middle ear and EAC on the right (Fig. 1). A subtotal petrosectomy with *en bloc* excision of the auricular pinna, EAC, timpanic membrane, malleus, incus, stapes suprastructure and omolateral functional lymph node neck dissection was performed. Through a retro-auricular incision a mastoidectomy was performed, the facial nerve was scheletonized from the stylomastoid foramen up to the geniculate ganglion and was found to be disease free. The incudostapedial joint was then disarticulated, EAC, in its osseous and cartilaginous parts, was completely isolated and removed in one piece together with the membrane and the ossicles, while the condyle of the mandible and parotid gland were free of disease and, therefore, preserved. The dura mater of the posterior and middle fossa were found to be infiltrated. The surgical cavity, having freed the dura from the visible tumour with electrocautery, was obliterated with abdominal fat and temporalis muscle flap. The cutaneous defect was covered with a parieto-temporal-occipital sliding skin flap.

Histologically, the lesion showed a variable mixture of morphologic patterns since tumour cells were arranged in tubular, trabecular, solid and cribriform nests or sheets. The cribriform spaces, in particular, often contained a variable amount of PAS-positive material. Occasionally, dense hyalinized collagenous stroma could be seen among the neoplastic nests. Moderate pleomorphism, areas of necrosis, many mitotic features (>5 per 10 high-power fields) were also detected in the tumour. Focal infiltration of skin and cartilage, by tumour cells, could be seen as well as neoplastic emboli in blood vessels.

Immunohistochemical findings (ABC method according to HSU) [19] revealed intense and diffuse positivity for cytokeratin, Vimentin (Fig. 2), muscle actin (Fig. 3) and focal staining for S-100 protein.

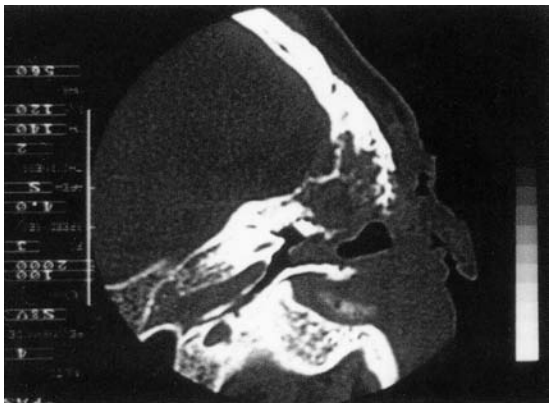

Fig. 1. High resolution CT scan showing lesion occupying external auditory canal.

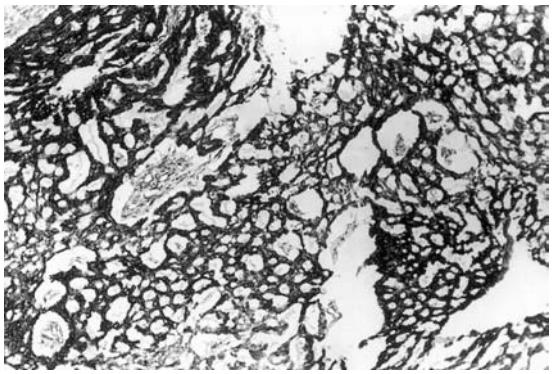

Fig. 2. Intense staining for Vimentin in neoplastic cells (avidin-biotin-peroxidase complex, X 100).

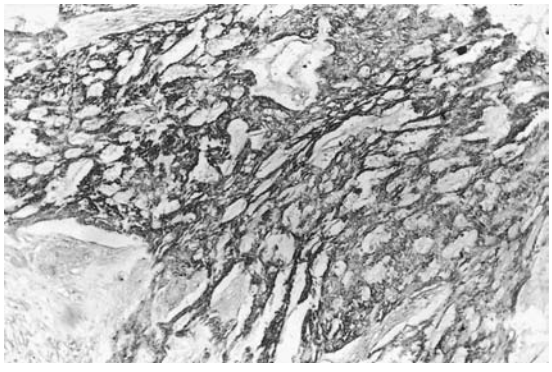

Fig. 3. Moderate staining for muscle actin in tumour cells (avidin-biotin-peroxidase complex, X 100).

Discussion

This case clearly demonstrates the aggressive local behaviour of ACC in this particular anatomical site. Indeed, the infiltration of the posterior wall of the EAC and the presence of neoplastic emboli within the vessels are clear signs of malignancy.
At the same time, since there was no evidence of parotid involvement, this case appears to be a primitive ACC of the EAC. Today, immunohistochemistry and scrupulous surgical planning with the aid of high resolution CT scans and magnetic resonance imaging have enabled a more accurate diagnosis to be made and, therefore, more radical treatment. Surgery is still the treatment of choice since radiotherapy has not been found to lead to a longer survival rate, but still has a role as a palliative measure or in combined therapy [15].

Conclusions

Surgery must be as radical as possible. The parotid gland and condyle of the mandible must be sacrificed if frozen sections reveal their involvement since local recurrences are very common and account for the high morbidity rate associated with this neoplasm. Metastases appear to be rare considering the long survival usually reported in these patients. For the same reasons, high suspicion and early diagnosis are still the best options to successfully treat this neoplasm.

References

1 Conley J, Schuller DE. *Malignancies of the ear*. Laryngoscope 1976;86:1147-63.

2 Hicks GW. *Tumors arising from the glandular structures of the external auditory canal*. Laryngoscope 1983;93:326-40.

3 Pulec JL. *Glandular tumors of the external auditory canal*. Laryngoscope 1977;87:1601-12.

4 Perzin KH, Gullane P, Conley J. *Adenoid cystic carcinoma involving the external auditory canal. A clinicopathologic study of 16 cases*. Cancer 1982;50:2873-83.

5 Anagnostou GD, Papademetriou DG, Segditsas TD. *Ceruminous gland tumors: report of three cases.* Laryngoscope 1974;84:438-43.

6 Batsakis JG, Hardy GC, Hishiyama RH. *Ceruminous gland tumors*. Arch Otolaryngol 1967;86:66-9.

7 Habib MA. *Ceruminoma in association with other sweat gland tumours*. J Laryngol Otol 1981;95:415-20.

8 Lynde CW, McLean DI, Wood WS. *Tumors of ceruminous glands.* J Am Acad Dermatol 1984;11:841-7.

9 Michel RG, Woodard BH, Shelburne JD, Bossen EH. *Ceruminous gland adenocarcinoma: a light and electron microscopic study*. Cancer 1978;41:545-53.

10 Nissim F, Czernobilsky B, Ostfeld E. *Hidradenoma papilliferum of the external auditory canal.* J Laryngol Otol 1981;95:843-8.

11 Ramadass T. *Cylindroma of external ear.* J Laryngol Otol 1966;80:863-6.

12 Turner HA, Carter H, Neptune WB. *Pulmonary metastases from ceruminous adenocarcinoma (cylindroma) of external auditory canal.* Cancer 1971;28:775-80.

13 Wassef M, Thomas V, Deffrennes D, Saint-Guily JL. *Primary adenoid cystic carcinoma of the skin. Histologic and ultrastructural study of two cases localized in the external auditory canal*. Ann Pathol 1995;15:150-5.

14 Umeno, H, Miyajima Y, Mori K, Nakashima T. *Clinicopathological study of 54 cases of adenoid cystic carcinoma in the head and neck.* Nippon Jibiinkoka Gakkai Kaiho 1997;100:1442-9.

15 Mills RG, Douglas-Jones T, Williams RG. *'Ceruminoma' – a defunct diagnosis.* J Laryngol Otol 1995;109:180-8.

16 Goodwin WJ, Yesse RH. *Malignant neoplasms of the external auditory canal and temporal bone.* Arch Otolaryngol 1980;106:675-9.

17 Hageman MEJ, Becker AE. *Intracranial invasion of a ceruminous gland tumor. A follow-up of 12 years.* Arch Otolaryngol 1974;100:395-7.

18 Mansour P, George MK, Pahor AL. *Ceruminous gland tumours: a reappraisal.* J Laryngol Otol 1992;106:727-32.

19 Hsu SM, Raine L, Fanger H. *Use of avidin-biotin-peroxidase complex (ABC) in immunoperoxidase techniques: a comparison between ABC and unlabeled antibody (PAP) procedures.* J Histochem Cytochem 1981;29:577-80.

■ Received: August 2, 2004
Accepted: October 5, 2004

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