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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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.

Introduction

Tumours of the external auditory canal (EAC) are extremely rare and only 20% of these are of glandular origin. The most frequent histotype is adenoid cystic carcinoma (ACC). Indeed, during the period 1928-1980, of 395,000 neoplasms examined at the Presbyterian Hospital in New York, only 25 appeared to be glandular tumours of the EAC and 10 of these were ACC. 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. 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.

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%). It is interesting to note that the origin of this tumour, following the hypothesis of Perzin et al., has been related to heterotopic salivary remnants rather than major salivary glands extension. 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) 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, indeed, several Authors suggest obtaining...
routinely frozen sections to evaluate local and perineural invasion. The 5-year survival rate is high 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, tympanic membrane, malleus, incus, stapes superstructure and omolateral functional lymph node neck dissection was performed. Through a retro-auricular incision a mastoidectomy was performed, the facial nerve was skeletonized 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) revealed intense and diffuse positivity for cytokeratin, Vimentin (Fig. 2), muscle actin (Fig. 3) and focal staining for S-100 protein.
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

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