CASE REPORT

Adenoid cystic carcinoma of external auditory canal: A case report

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Abstract

Primary malignancies of the external auditory canal (EAC) are extremely rare with more than 80% being squamous cell carcinomas and adenoid cystic carcinoma (ACC) accounting for approximately 5%. These tumours are associated with a high risk of recurrence and significant morbidities from surgical management and adjuvant radiotherapy. Therefore, we present a case of young female with right EAC mass diagnosed as cylindroma and confirmed postoperatively to be ACC of the EAC which is a rare case in the literature.

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1. Introduction

Adenoid cystic carcinoma (ACC) is a rare epithelial tumour entity and comprises about 1% of all malignant tumours of the oral and maxillofacial region. ACC, first described as “cylindroma” by Billroth, is commonly classified with the salivary gland tumours, although it may arise in any site where mucous glands exist. Half of these tumours occur in glandular areas other than the major salivary glands, principally in the hard palate, but they also arise in the tongue and in other areas that are the sites of minor salivary glands. Unusual locations include the external auditory canal, nasopharynx, lacrimal glands, breast, vulva, oesophagus, cervix, and Cowper glands.

The long natural history of this tumour, its propensity for perineural invasion, and its tendency for local recurrence are well known. Although it presents a widespread age distribution, peak incidence occurs predominantly among women, between the 5th and 6th decades of life. It is a slow growing but highly invasive cancer with a high recurrence rate. Lymphatic spread to the local lymph nodes is rare. Haematogenous spread, however, occurs often in the course of the disease. Perineural spread of ACC has long been recognized. The literature revealed the region of Gasserian ganglion to be the most common site of involvement (35.8%). Microscopically, it is...
The patient then defaulted on the treatment till June 2011 when she claimed that the swelling increased in size with intermittent pus discharge, but with no postauricular swelling or pain. No nausea or vomiting, however headache was predominant. On examination there was protruding mass from the right external auditory canal that increased in size as compared to before. Repeated HRCT temporal bone (Fig. 2) showed ill-defined enhancing mass arising from the right pinna was now larger measuring 3.7 cm × 1.9 cm that occupies the entire right EAC into the right middle ear cavity. Superiorly, it invades into the right mastoid air cell with more destruction of the septa. There was more break of the floor of the right temporal bone. The right scutum complex eroded and had no evidence of intracranial extension.

The right modified radical mastoidectomy was performed on 15/8/2011. Intraoperative findings were large EAC granuloma with stalks arising from anteroinferior and part of the posterior wall of EAC. The ossicles were surrounded by the granulation tissue which also filled the middle ear. As there was no facial nerve monitoring in our centre, the facial nerve was not properly traced and identified. Grossly, facial canal seems to be intact. No pus or keratin noted. Mastoid cavity looks sclerosed.

Postoperatively, the patient has immediate facial nerve palsy (House Brackman Grade III) due to intraoperative injury. Histopathological examination confirmed it as adenoid cystic carcinoma. Magnetic resonance imaging (MRI) of the internal auditory meatus and brain (Fig. 3) was performed and showed features that represent tumour residual with local extension, however no obvious cerebellopontine angle or brain parenchymal lesion. Then, the patient was referred to Kuala Lumpur General Hospital (tertiary hospital) for further management. Tumour debulking surgery was performed. Intraoperative findings were tumour mass occupying mastoid antrum and middle ear adhering to the tympanic membrane and also extending into the Eustachian tube. The patient was then referred to oncology for their expert opinion and management and completed her radiotherapy session there.

3. Discussion

Malignant tumours of the EAC are rare and most are squamous cell carcinomas. ACC arising in the EAC is exceedingly rare. Although ACC is a rare EAC tumour, it is relatively common in the salivary glands of the head and neck. ACC growth rate is slow and the nature of this carcinoma shows a slow malignant course. The true origin of ACC in the EAC is controversial. It has been proposed that these tumours arise from the ceruminous glands. Microscopic studies of these tumours and ceruminous glands demonstrate similar histologic features. Some authors have suggested that these tumours arise from the ectopic salivary glands of the EAC, although this opinion has not been proved. In general, ceruminous gland tumours can be classified as ceruminous adenoma, pleomorphic adenoma, ceruminous adenocarcinoma and adenoid cystic carcinoma. ACC has 3 main histological patterns: tubular, cribriform and solid.10,11 In the salivary glands, the prognosis of ACC correlates with the predominant histological pattern. Tubular ACC has the best prognosis, whereas solid ACC has the worst prognosis.11 However, in the EAC there was no significant correlation between these histological patterns.
Some factors including tumour positivity at surgical margins, parotid gland and adjacent bone involvement, perineural invasion and local recurrences are associated with aggressiveness and high mortality rate. When relapse develops, it generally occurs within two years. Late relapses were rarely reported as much as 14 years. In some of the cases, pulmonary metastasis was detected 20 years after treatment; this is the longest period for metastases in the literature for adenoid cystic carcinoma. Metastases into the lung are more common than regional lymph node metastases. Rarely widespread visceral involvement may be seen.

The performance status of the patient and metastasis are important factors for selecting the treatment modality. The survival with surgery and postoperative radiotherapy is better than that with surgery alone. The presence of tumour free surgical margins can be correlated with a better local control and also with longer survival. Treatment consisted of radical excision of the EAC via a modified temporal bone resection. In tumours which were thought to be aggressive (invaded to the adjacent tissue, higher grade or impossible to get negative surgical margins) elective neck lymph node dissection could be added to surgery. Radiotherapy and chemotherapy are not curative but can help in palliation and as adjuvant therapy.

References