Rare disease

Adenoid cystic carcinoma of base of tongue

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Adenoid cystic carcinoma (ACC) is a rare malignant tumour arising from salivary glands. ACC of the head and neck shows a long natural course, high recurrence rates, late metastasis and a tendency for perineural invasion. The authors present a woman with ACC at base of tongue with good response to radiotherapy.

BACKGROUND

Adenoid cystic carcinoma (ACC) is a rare malignant tumour accounting for less than 1% of all head and neck malignancies.¹ Most ACCs arise in the minor salivary glands (60%). ACC of minor salivary gland origin occurs most frequently in the oral cavity (palate).² The frequency reported in the tongue is variable, between from only 1% to 20.8%.³ Majority of these tumours are located in the base of tongue.⁴⁻⁵ Clinically, it is characterised as a painless slow growing mass with a high recurrence rate and metastasis to other organs.⁴ The prognostic factors consist of stage of disease, location of tumour, solid growth pattern, size of lesion, state of surgical margins and intraneural and/or perineural invasion.²⁻⁴⁻⁶ The choice of therapy is influenced by site, stage, histologic grade and biologic behaviour of the ACC.² Although in most cases surgical resection with negative margins is the cornerstone of treatment⁵⁻⁷ postoperative radiotherapy is often recommended because of infiltrative growth pattern, and presence of perineural invasion.¹⁻³⁻⁶⁻⁷ Some authors suggest that advanced and non-resectable tumours may be treated only with radiotherapy.⁵ In this article, we present a 59-year-old woman with ACC at the base of tongue.

CASE PRESENTATION

A 59-year-old woman with dysphagia and feeling of discomfort in oral cavity referred to a private ear, nose, throat (ENT) clinic in Yazd, Iran. The ENT specialist following examination diagnosed a mass at the patient's base of tongue and biopsy confirmed ACC. The patient was operated at Tehran, capital of Iran, therefore we do not have detailed information on her surgical procedure; and then referred to our Centre Milad Hospital in Isfahan (the only centre in Iran equipped with linear accelerator with multi-leaf collimator) for adjuvant radiation therapy. Pathology report showed a 2×2 ACC tumour in the central part of base of tongue with perineural and positive surgical margins. MRI showed gross tumour residue (figure 1); but other findings were normal. Postsurgical examination did not reveal any reconstruction evidence.

DIFFERENTIAL DIAGNOSIS

Pleomorphic adenoma with cribriform pattern, polymorphous low-grade adenocarcinoma and basaloid squamous cell carcinoma should be considered in the differential diagnosis.

TREATMENT

Because of perineural invasion, the radiation field was extended to the base of skull. Although there is no any agreement for elective lymph node treatment we decided to treat neck with a low neck anterior field with a 6 MV photon energy to 4500 cGy. Then we excluded the spinal cord from the treating field and increased the dose of radiation to tumour and to base of skull to 6000 cGy. Finally, we

Figure 1 MRI showed gross tumour residue.

Figure 2 Repeated MRI 4 weeks later showed very good response to radiation.

completed the treatment with a 400 cGy boost on tumour residue. The patient tolerated the treatment fairly well and only mild degree of mucositis happened and she lost two kilograms weight.

OUTCOME AND FOLLOW-UP

Repeated MRI 4 weeks later showed very good response to radiation (figure 2). One year after treatment she is well without any evidence of relapse.

DISCUSSION

ACC is a rare epithelial tumour with a paradoxical behaviour. Although indolent local recurrences is common due to extensive local tissue infiltration and perineural spread, despite aggressive surgical resection. Metastatic spread to regional lymph nodes is uncommon, but distant metastasis to the lungs and bones is frequent. Despite metastasis long-term survival is common.^{2 5 7} Many of authors believe that the base of tongue is the second most common site of intraoral ACC after hard palate, however with a wide range of incidence.^{3 5} On the other hand, according to a study performed by Roper *et al* in University of Texas ACC is the most frequent histology in the base of tongue.⁸ As we explained above, treatment of this tumour in this site is more difficult. Surgery is an essential part of treatment for almost all of patients; however it remains to decide which of the radical or conservative surgeries should be preferred? Most investigators such as Mclean *et al*¹ have demonstrated that tumours with positive surgical margins have a worse prognosis. Positive surgical margins may be a result of tumour aggressiveness not inadequate surgical procedure. Most specialists believe that adding postoperative radiotherapy decreases the local recurrence, but: 'Dose it lead to prolonged survival?' maybe not. However,

according to report of Miglianico *et al*. One hundred and two patients with cervico-facial ACC were treated with surgery alone, radiotherapy alone or both. There was no significant difference in the NED 5-year survival rate according to sites or treatments.⁹

The lack of survival advantage for patients treated with combination surgery and radiotherapy is assumed to be due to the high rate of distant metastases and the relatively high likelihood of long-term survival after salvage therapy for patients who experienced locoregional recurrence.¹⁰ Because of characteristics of ACC, postoperative radiation therapy is the treatment policy employed in most centres, but what could be the adequate radiation dose? May be there is a dose dependent radiation policy for local control of ACC; but a wide range was advised: from >45 Gy as shown by Vikram *et al* and >56 Gy, with statistical significance attained for patients with positive margins as described by Garden *et al* and finally >60 Gy as shown by Simpson *et al*. In the study performed by Hsuan-Chih Hsu *et al*, however there was not seen any statistical difference in overall survival rates, local control rates and distant metastasis-free rates for patients receiving ≥63 Gy and <63 Gy.⁷ There are no definite agreements about the irradiated fields for ACC of the head and neck, but the base of the skull is routinely included in the postoperative treatment volume when there is perineural invasion. Another question is about elective neck irradiation. Although the risk of occult metastases is low, Mendenhall *et al* advised to electively treat the first-echelon nodes for patients with primary tumours in sites rich in capillary lymphatics, such as the base of tongue and nasopharynx.⁹ According to this information we treated patient with 6400cGy and extended field to base of skull and treated the neck prophylactically. Repeated MRI 4 weeks after termination of treatment revealed very good response. Six months follow-up indicated that the disease was controlled.

Learning points

- ▶ ACC is a rare tumour of head and neck and is a challenging treatment issue.
- ▶ Radiation therapy can be effective despite gross residue.
- ▶ High dose radiation (more than 60 GY) is essential for effective treatment.

Competing interests None.

Patient consent Obtained.

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