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Case report

Angiosarcoma of the auricle

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ABSTRACT

Introduction: Angiosarcomas are rare but aggressive vascular cancers of endothelial cell origin. The diagnosis is often difficult, based on specific immunohistological features. They characteristically present a high local recurrence rate and an early metastatic potential.
Case report: We report a case of angiosarcoma of the auricle in a 31-year-old female patient who presented with swelling of the left ear for 3 years. Histological examination demonstrated angiosarcoma. Treatment consisted of surgery followed by postoperative radiotherapy.
Discussion: This case report describes the epidemiological and clinical profile and the treatment and outcome of angiosarcoma of the auricle, together with a review of the literature, demonstrating certain diagnostic and therapeutic difficulties in the management of this aggressive tumour.

1. Introduction

Sarcomas are malignant mesenchymal tumours, representing less than 1% of all cancers [1]. Angiosarcomas represent 2% of all sarcomas and arise from vascular endothelial cells [2]. However, 50% of all angiosarcomas involve the skin of the head and neck [3]. The diagnosis of this tumour is often delayed by its apparently benign clinical presentation [4], which can be confused with a skin infection or soft tissue trauma [4]. The auricle is an exceptional site of angiosarcoma, as, to our knowledge, only 4 cases have been reported in the literature [5].

2. Case report

A 31-year-old woman, with no notable history, presented with progressive swelling of the left ear for 1 year in a context of good general health. A skin infection was initially suspected and the patient was treated with antibiotics for three months, and was subsequently lost to follow-up for 9 months. Clinical examination revealed a painless spherical lesion in the pinna of the left ear measuring 4 cm × 4 cm, that bled on palpation, with no facial palsy or cervical lymph nodes and with normal otoscopy (Fig. 1).

Computed tomography showed a soft tissue tumour invading the left auricle and adjacent skin (Fig. 2), with no underlying contrast-enhanced bone lesion.

Biopsy of the lesion and the left pre-auricular skin revealed a diagnosis of angiosarcoma. Immunohistochemistry revealed the following results: vimentin (+), cytokeratin (−) and S-100 (−). Treatment consisted of complete surgical resection of the mass with superficial parotidectomy and systematic levels I, II and III lymph node dissection (Fig. 3). Histological examination of the operative specimen demonstrated moderately well differentiated high-grade angiosarcoma invading muscle and cartilage with negative lymph nodes N0 R−. External beam radiotherapy was delivered to the tumour bed at a dose of 50 Gy. After 6 months of follow-up, this patient was in good general health with no signs of recurrence.

3. Discussion

Angiosarcoma is one of the rarest soft tissue tumours; it is very aggressive and has a poor prognosis [1]. It is a very well differentiated tumour that can be confused with anaplastic tumours and haemangiomas, which are difficult to distinguish from carcinomas [3]. Angiosarcoma of the auricle is extremely rare, as, to our knowledge, only 4 cases have been reported in the literature [1–5]. It can be considered to be an angiosarcoma of the face and scalp [3]. It has a poor prognosis, with a 5-year survival rate of 12% [6]. The initial clinical presentation of the tumour is extremely variable. The patient often presents late, as the swelling is rarely painful [3], and may simply resemble a bruise or an ulcerated and infected nodular tumour. At a more advanced stage, the tumour is typically haemorrhagic and ulcerated [7] and may resemble a benign soft tissue tumour such as haemangioma or malignant skin carcinoma [4]. This initial clinical confusion can lead to delayed diagnosis and may predispose to tumour progression, while tumour size constitutes an

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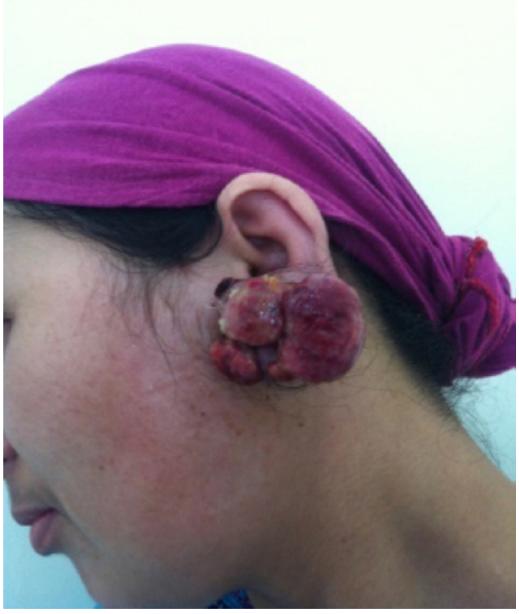

Fig. 1. Swelling of the auricle of the left ear.

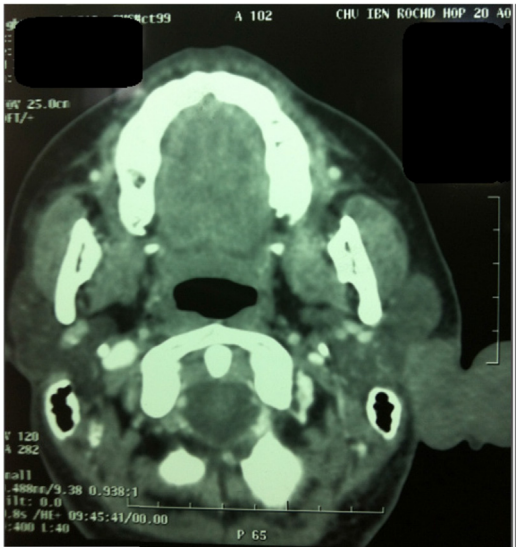

Fig. 2. CT scan showing invasion of the soft tissues of the left auricle.

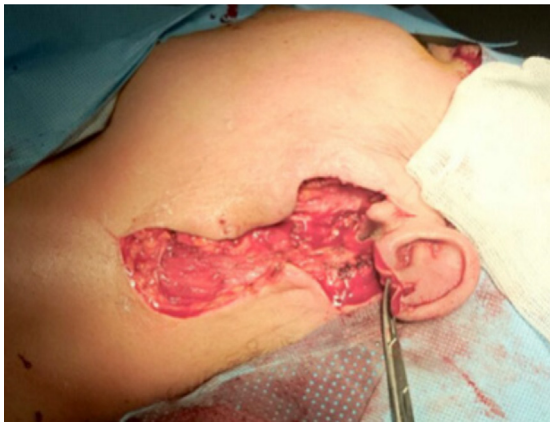

Fig. 3. Resection of the mass with superficial parotidectomy and levels I, II and III lymph node dissection.

important prognostic factor. In the series reported by Lydiatt et al., all patients with an angiosarcoma larger than 10 cm died, while smaller lesions measuring 5 cm were associated with a higher survival rate, hence the value of early diagnosis [8]. Only histological examination can confirm the diagnosis [5]. Histologically, angiosarcoma is composed of a network of vascular anastomoses or a disorganized sinusoidal appearance dissecting the dermal collagen [7]. Computed tomography is useful to evaluate the tumour architecture and characteristics and for precise staging of the tumour [5] with invasion of the soft tissues of the ear and the adjacent skin [9].

In the absence of consensus, the Pittsburgh TNM classification of squamous cell carcinomas of the external auditory canal [9,10], proposed in 1990, is the most commonly used classification, based on the findings of preoperative clinical examination and CT scan [8]. The advantage of this classification is that it can be used to guide both treatment and prognosis. A minor revision was proposed in 2000 [8].

Distant metastases may involve cervical lymph nodes, as well as liver, spleen, bone, kidneys and myocardium [6]. Surgical resection with large and deep resection margins is recommended [7]. In the case reported here, and after multidisciplinary consultation, it was decided to perform levels I, II and II lymph node dissection in addition to large surgical resection, in view of the high incidence of lymph node metastases (10–15%) [6].

The role of adjuvant radiotherapy remains controversial, as no study had demonstrated any benefit on survival [3]. In the presence of cervical lymph node metastases, Hodgkinson et al. recommended lymph node dissection and radiotherapy to the tumour bed and to the neck [10].

These tumours have a poor prognosis due to their tendency to local recurrence and lymphatic or haematogenous metastases. Lydiatt et al. reported a 5-year survival rate of 33%, with a recurrence rate of 78% [8]. Our patient did not present any signs of recurrence 6 months after the end of treatment.

4. Conclusion

Angiosarcoma of the auricle is a rare tumour. The diagnosis is delayed due to its nonspecific early signs. Histological examination of a biopsy, performed in the case of any suspicious lesions, is the key to the diagnosis and the only way to ensure effective treatment. Multidisciplinary management by a surgeon, radiologist and radiotherapist is essential.

Disclosure of interest

The authors declare that they have no competing interest.

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