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Clinical Records

Haemangiopericytoma of the middle ear with benign histological features

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Abstract
Haemangiopericytoma is a rare vascular tumour, particularly in the head and neck region. We described the first case of haemangiopericytoma arising from the middle ear in 1995. The present case is the second example of a primary middle-ear haemangiopericytoma with benign histological features. Clinical photography, computed tomography (CT) and magnetic resonance imaging (MRI) scans and histological photographs depict the findings.

Key words: Haemangiopericytoma; Temporal bone; Ear, neoplasms

Introduction

Haemangiopericytoma, an uncommon vascular tumour, was first described by Stout and Murray (1942) as a tumour consisting of proliferating pericytes that surround capillaries. Since pericytes surround all capillaries, haemangiopericytoma can be found in any part of the body. In the head and neck region, haemangiopericytoma originates usually from the sinonasal tract, nasopharynx, maxilla and the mandible. Skull base, oral cavity, infra temporal fossa and orbital haemangiopericytoma cases have also been reported. Only eight temporal bone haemangiopericytomas have previously been reported (Sutbeyaz, *et al.*, 1985) and in the most recent literature review, a case of a temporal bone haemangiopericytoma with no evidence of middle-ear involvement (Cross and Mixon, 1996) has also been reported. However, there is only one reported case of haemangiopericytoma arising from the middle ear (Sutbeyaz *et al.*, 1995). In this article the second example of primary middle-ear haemangiopericytoma is presented.

Case report

A 60-year-old female was referred to us with suppurative ear discharge, hearing loss, and a mass in the right external auditory canal, and also a history of otorrhagia, from time to time, during the previous five years. Examination showed a pinkish-grey polypoid mass, which totally filled the right external auditory canal, associated with purulent ear discharge. There was no palpable lymph node or metastatic disease. Computed tomography (CT) scan of the temporal bone showed a soft tissue mass, that filled and enlarged the right middle-ear cavity and right external auditory canal (Figure 1). Magnetic resonance T2-weighted and gadolinium-enhanced magnetic resonance (Figure 2) images were obtained and demonstrated the mass showing high signal intensity in the right middle-ear cavity. Pure tone audiometric measurement revealed right-sided profound conductive hearing loss.

With retroauricular incision, right middle ear and mastoid cavities were explored. A polypoid mass filling the right middle-ear cavity, external auditory canal and mastoid antrum was observed. The mass was attached to adjacent middle ear and mastoid structures. The middle-ear cavity and mastoid antrum were expanded by compression of the mass. A radical mastoidectomy was performed and the mass was detached from the bony structure, and removed. Excessive bleeding was controlled.

The gross appearance of the specimen was grey-tan with a soft rubbery consistency. Light microscopy showed that

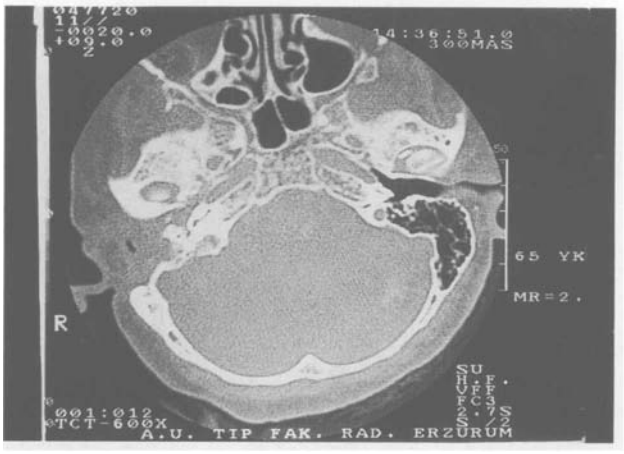

Fig. 1

CT scan showing an enlargement of the right middle-ear cavity by the soft tissue mass that totally fills the middle ear and external auditory canal.

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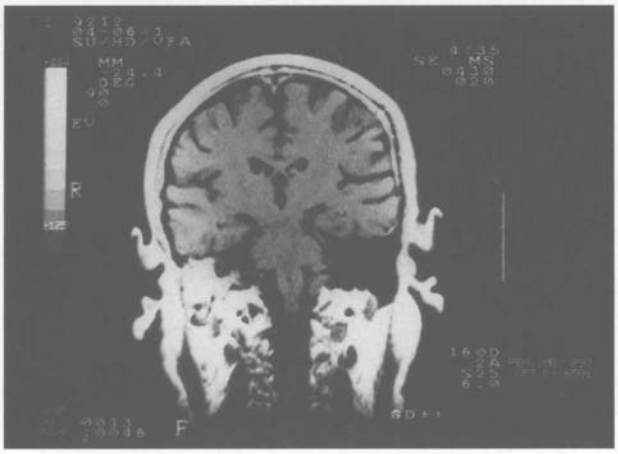

FIG. 2

Gadolinium-enhanced magnetic resonance image showing the well-defined mass with high signal intensity in the right middle-ear cavity.

the tumour was covered by squamous epithelium of the drum remnant and also showed round and spindle-shaped cells with ill-defined borders and round to oval nuclei surrounding the endothelial-lined vascular channels (Figure 3). Mitoses, cellular pleomorphism, necrosis and haemorrhage were not seen. A reticulin preparation revealed the dense reticulin meshwork surrounding vessels and tumour cells. A diagnosis of benign haemangiopericytoma was made. Immunostaining for neuron-specific enolase (NSE), chromogranin, factor VIII-related antigen, S-100 protein and for vimentin were performed in order to confirm the diagnosis. The tumour cells were negative for NSE, chromogranin, S-100 and factor VIII-related antigen, which is positive for endothelial cells, but positive for vimentin (Figure 4).

Eight months later, the patent was healthy with the exception of the right-sided profound conductive hearing loss and had no evidence of recurrent tumour.

Discussion

Haemangiopericytoma is a rare vascular tumour, existing anywhere in the body but commonly found in the soft tissues of the trunk and lower extremities. The tumour may appear at any age with equal male-female distribution, but the great majority of patients are older than 20 years

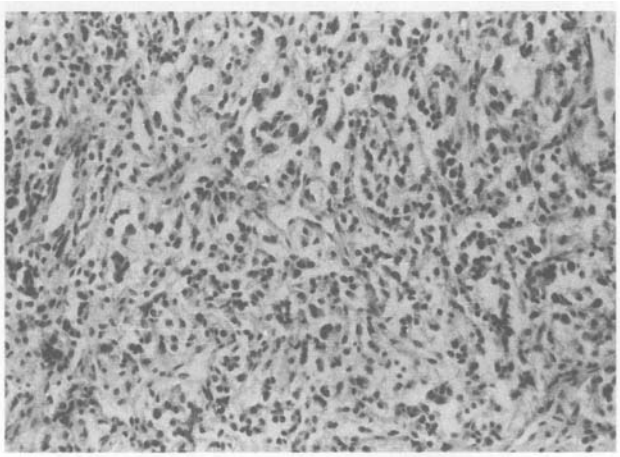

FIG. 3

Histological appearance of haemangiopericytoma. Section of the tumour showing irregular vascular channels surrounded by round and spindle-shaped cells (H&E; ×200).

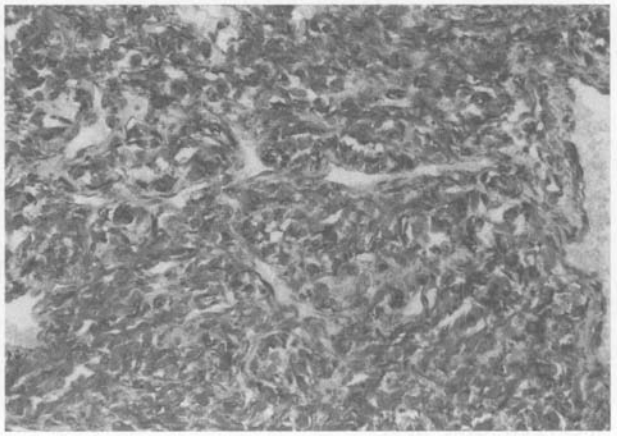

FIG. 4

Positive immunohistochemical staining of tumour for vimentin (×100).

(Sabini, *et al.*, 1998). It represents one per cent of all vasoformative neoplasms, and 15 to 30 per cent of haemangiopericytomas occur in the head and neck region (Güdrün, 1979; Batsakis and Rice, 1981; Sabini *et al.*, 1998). The majority of the reported cases are in the nasal cavity, paranasal sinuses, nasopharynx, maxilla, mandible and the orbit (Enzinger and Smith, 1976; Batsakis and Rice, 1981).

In our previous middle-ear haemangiopericytoma case report, a review of the literature revealed only seven cases of temporal bone haemangiopericytoma without any primary middle ear involvement, and the majority of these were malignant (Sutbeyaz *et al.*, 1995). We reviewed the most recent literature. There is one more report of a temporal bone haemangiopericytoma case. Cross and Mixon (1996) presented a benign haemangiopericytoma in the mastoid antrum with no evidence of middle ear involvement.

McMaster *et al.* (1975) have categorized haemangiopericytomas histologically as: benign (low-grade); histologically borderline (intermediate grade); and histologically malignant (high-grade). They expected malignant behaviour in cases having a slight degree of cellular anaplasia or one mitotic figure per 10 high-power fields or having a moderate degree of cellular anaplasia and one mitotic figure per 20 high-power fields. Enzinger and Smith (1976) emphasized that four or more mitotic figures per 10 high-power fields were indicative of a rapidly growing tumour capable of recurrence and metastasis. They also suggested that malignant haemangiopericytomas tend to be more cellular with cellular pleomorphism, necrosis and haemorrhage. Although these categories exist, it has been known that this tumour has a rather unpredictable behaviour even if it is thought to be benign histologically. Histologically, haemangioma, haemangioepithelioma and glomus tumour can be confused with the benign form. The pericytes, first described by Zimmerman (1923), are the contractile cells surrounding the capillaries, which are round or spindle-shaped with hyperchromatic nuclei. Proliferation of these cells surrounding the capillaries characterizes the haemangiopericytoma (McMaster *et al.*, 1975; Güdrün, 1979; Birzgalis *et al.*, 1990).

Modern imaging techniques such as CT scan, ultrasound, Doppler sonography arteriography, and MRI can give information about the precise localization and the extent of tumour invasion into the surrounding tissue. Recent studies emphasized that in head and neck haemangiopericytoma cases, MRI should include determi-

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nation of the extent of the tumour. T2-weighted images and gadolinium-enhanced images disclose a well-defined tumour as a hyperintense mass (Tanaka *et al.*, 1996; Sabini *et al.*, 1998).

The differential diagnosis of the benign haemangiopericytoma in the middle ear includes other spindle cell mesenchymal tumours such as fibrous histiocytoma, that can mimic haemangiopericytoma, and especially paraganglioma, that is the most common neoplasm of the middle ear (Porter *et al.*, 1991). Immunohistochemistry is useful for the differential diagnosis. Haemangiopericytoma is positive for vimentin and CD34 and negative for S-100 protein, NSE, chromogranin (Middleton *et al.*, 1998). Paragangliomas are positive for S-100 protein, NSE, chromogranin but negative for vimentin (Gupta *et al.*, 1998). Vimentin and CD34 staining are also characteristic of fibrous histiocytoma (Dalley, 1999). Staining for factor VIII-related antigen is positive in the endothelial cells of the capillaries, but negative in the tumour cells of haemangiopericytoma, it is negative for fibrous histiocytoma.

Optimal treatment of haemangiopericytoma is complete surgical excision. The recurrence rate of the tumour is relatively high (McMaster *et al.*, 1975). It has been reported that pre-operative embolization may be helpful in order to obtain a better surgical result (Birzgalis *et al.*, 1990). Although the tumour seems to be radioresistant, radiotherapy may be useful after incomplete resection of the malignant form. The role of chemotherapy in the treatment of the malignant haemangiopericytoma is still obscure. A benign form of the haemangiopericytoma does not exclude the possibility of recurrence; therefore close long-term follow-up is mandatory.

Haemangiopericytoma of the temporal bone is extremely rare. Although nine cases of temporal bone haemangiopericytoma have been presented, only one of them had middle ear involvement. The present case of benign haemangiopericytoma is therefore the second case primarily originating from the middle ear.

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