CASE REPORT

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Haemangiopericytoma in the distal third of the arm

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Abstract A 49-year-old woman had a haemangiopericytoma in the distal third of the arm, which is an extremely rare location. There was no recurrence of the tumor 5 years after wide margin surgical excision.

Résumé Présentation d'un cas d'hémangiopéricytome du tiers distal du bras chez une femme de 49 ans. Une résection large a été faite. Il n'y a pas de récurrence au recul de 5 ans.

Introduction

Haemangiopericytoma is an unusual vascular tumor, which arises from the pericapillary pericytes cells of Zimmermann. Such tumors have been described in the lower legs, the pelvis and the retroperitoneal area. Uncommon cases have been reported involving the trunk, larynx, spleen and central nervous system. We describe here a case of haemangiopericytoma in the distal third of the arm.

Case report

In March 1994, a 49-year-old woman was referred to us complaining of severe pain and swelling in her right arm. The swelling itself had been noted 10 years prior to her visit. A firm, tender, roundish mass 12 cm in diameter was found arising from the radial aspect of the distal arm, and radiographs showed a soft oval mass with an isolated central calcification. Magnetic resonance images (MRI) before and after administration of gadolinium showed a well-defined tumor beneath the bellies of the biceps brachii, anterior brachialis, brachioradialis and the long and short extensor muscles of the wrist (Fig. 1). Angiography demonstrated extensive tumor neovascularity (Fig. 2).

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Fig 1 Magnetic resonance image (MRI) showing the ovular mass underneath the anterolateral muscles of the right arm

Open biopsy showed the lesion to be a grade 1 haemangiopericytoma. The patient received adjuvant radiotherapy but no chemotherapy. A wide margin excision of the tumor was carried out, and the gross specimen consisted of a greyish mass, 8 x 6 cm, with no areas of hemorrhage or necrosis. Histological and histochemical examination revealed a mass composed of well-delineated perivascular nodules of spindle-shaped tumor cells (Fig. 3). The mitotic index was 0–1 high power fields. The tumor was negative for acid protein s-100 and focally positive for alpha smooth muscle actin. Five years later there was no recurrence of the tumor.

Discussion

The term "haemangiopericytoma" was first coined by Murray and Stout [10] in 1942 to describe a series of un-

Fig. 2 Angiography demonstrating the tumor richly vascularised by the humeral artery

common neoplasms related to glomus tumors. In 1968, Murad and Haam [9] demonstrated that haemangiopericytoma and glomus tumors are histologically distinct entities, the former arising from pericytes and the latter from glomus cells. Because of the ubiquity of pericytes in mesenchymal tissues, haemangiopericytomas may occur in any part of the body. Nevertheless, the occurrence of this tumor in the upper limb has been reported previously in only two cases [4, 6].

Fig. 3 The typical histological appearance of haemangiopericytoma showing slit-like vascular spaces surrounded by spindle-shaped cells ($\times 150$, haematoxylin and eosin)

The growth of haemangiopericytomas is generally slow: pain may only appear months or years after the onset and is caused by the compression of surrounding nerves [1]. Predicting aggressive tumor behavior in haemangiopericytomas has proved to be very difficult. Several authors have reported an association of mitotic activity, necrosis and hemorrhage with more malignant tumors [1, 2, 7].

Considering the mitotic activity, the absence of hemorrhagic and necrotic areas and the location, a wide margin resection was the treatment of choice for our patient. With such a resection recurrence is rare, and this is confirmed by several authors [1–3, 7]. Chemotherapy was not administered because it has not been shown to be useful in the management of resectable haemangiopericytomas [1, 11]. On the other hand, adjuvant radiotherapy was given because this is considered to be effective in local management [1, 5, 8] and should be included along with resection in the treatment regimen of these tumors.

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