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Hemangiopericytoma in the Mental Region: A Case Report

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Abstract: ***Hemangiopericitoma is a rare tumor that originates from the pericytes. Only histology permits a reliable diagnosis. There are no known parameters to predict the biological behavior of the tumor, so every hemangiopericitoma has to be treated as potentially malignant. No age or gender prevalence of this tumor in the region of the head neck has yet been observed. Hemangiopericitoma should be treated by radical surgery; chemotherapy or radiation should be reserved for incompletely removed or metastatic tumors. Metastasis and recurrences have been described even decades after first tumor treatment, so that all patients should undergo life-long follow-up.***

Keywords: hemangiopericitoma, tumour, maxillofacial

1. Introduction

Hemangiopericytoma is an uncommon vascular neoplasm that originates from small pericapillary situated spindle-shaped cells called Zimmermann's pericytes. First described by Stout and Murray in 1942, hemangiopericytoma accounts for no more than 1% of all vascular tumors (1). Because pericytes are located around capillaries, hemangiopericytoma can be found in any part of the body. They are most common in the lower extremities, the pelvis, and the retroperitoneum. Approximately 15% occur in the head and neck region (6) with lesions reported arising in the lip, tongue, maxilla, mandible, floor of mouth, buccal mucosa, retromolar, gingival, laryngeal and parapharyngeal spaces (3).

In literature there are distinguishing signs between the benign and the malignant forms. Indicative for a malignant transformation are the increased mitotic activity, higher cell density, the appearance of non-differentiated cells as well as necrotic and hemorrhagic zones in the tumour tissue (4). Clinically, the malignant hemangiopericytoma is characterized by haematogenic metastases, typically in the lung, the liver, and the skeletal system. On first examination it is generally difficult to distinguish between benign and malignant forms. A more recent investigation tried to define the differences between the two forms using a molecular-biological marker (Ki-67) to calculate the proliferation index (5). In some cases, the malignant nature of the hemangiopericytoma is established only when the tumour recurs or metastasizes.

The tumour may occur in any age, although the peak incidence is in the fifth and sixth decades. There is an equal sex distribution (6). Pain is an uncommon feature and relates to visceral or neural compression by the tumour (6).

The diagnosis of this tumour is problematic. The physical examination often only confirms that the patient has a tumour (6). Laboratory data and imaging modalities including plain radiography, angiography, computed tomography, and magnetic resonance imaging are not helpful in confirming a definitive diagnosis, which is made only on the basis of the histological examination (1).

Macroscopically hemangiopericytoma is usually light gray to dark tan and is occasionally interrupted by areas of hemorrhage or necrosis. The tumour is described as elastic, hard, and solid mass and, despite their malignant behaviour, the neoplasm are frequently noted to be encapsulated (7).

Surgery is the standard therapy when the tumor is localized and technically resectable. Radiation therapy has usually been preserved for unresectable and recurrent tumors (6). Postoperative radiation therapy, with a dose of approximately 50 Gray or higher, has been recommended to reduce the rate of local reccurence (6). The literature does not provide sufficient evidence of the effectiveness of chemotherapy (7). Craven et al. (6) have recommended routine angiography and preoperative embolization before wide excision because hemangiopericytoma is usually a highly vascularized tumour.

In stating the differential diagnosis, care must be taken to distinguish hemangiopericytoma from meningeoma and the mesenchymal chondrosarcoma (1).

We present the case of a 64-year old man with a 10-year history of a gradually enlarging swelling in the left mental region (fig. 1). In the last 2 years the tumour has grown significantly. Clinical examination revealed a nodular lesion, 5 cm in diameter, which was covered by skin with teleangiectasies.(fig. 2) The lesion was not fixed to the adherent tissues.(fig. 3) No neck lymphadenopathy could be detected. After general evaluation, the surgical exploration was performed. Good exposure was achieved at the price of minor aesthetic and functional impairment. On the basis of the histological and immunohistochemical findings, the final diagnosis was hemangiopericytoma. The resected surgical margins were free of tumour cells. Radiation therapy was administered. No signs of local recurrence or distant metastasis have been observed.

2. Conclusion

Malignant hemangiopericytoma continues to be a diagnostic, therapeutic and prognostic challenge. Adequate therapy and life-long follow-up are therefore mandatory in haemangiopericytomas. Complete surgical resection of the

tumour is usually the treatment of choice, in spite of the high degree of surgical skill required, especially in advanced tumour and in case of recurrence.

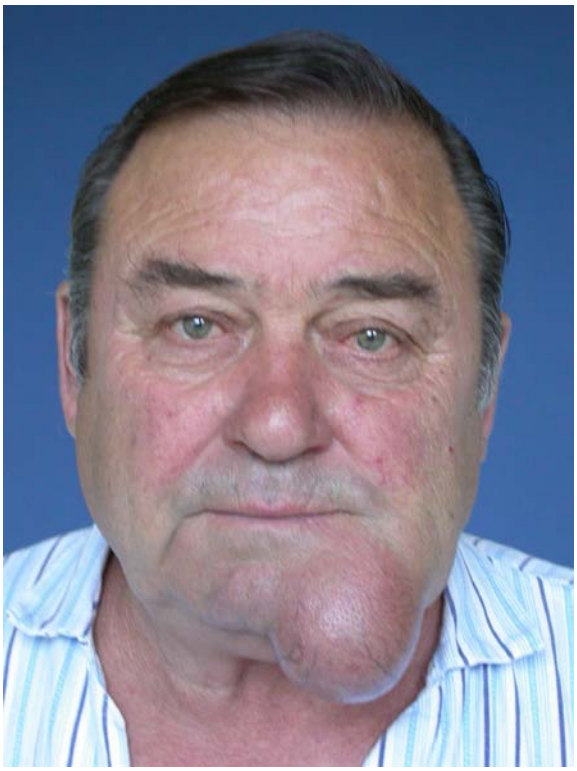

Figure 1: Hemangiopericitoma in mental region

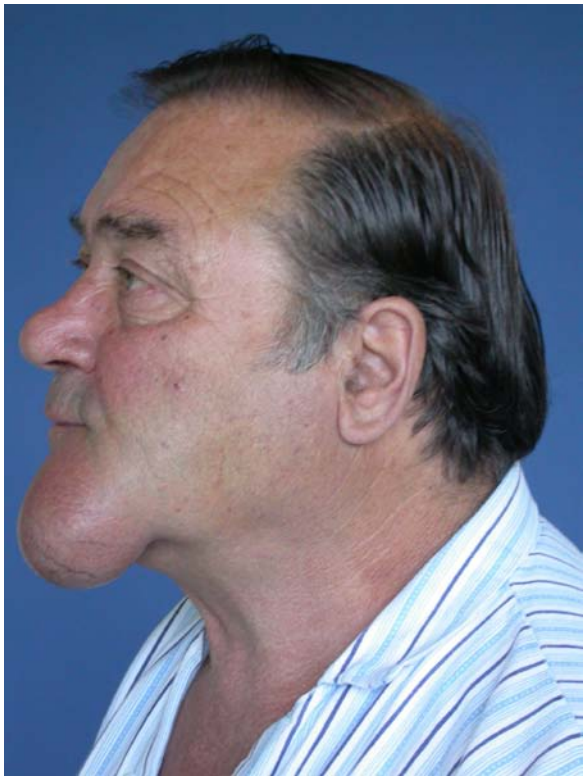

Figure 2: Hemangiopericitoma in mental region

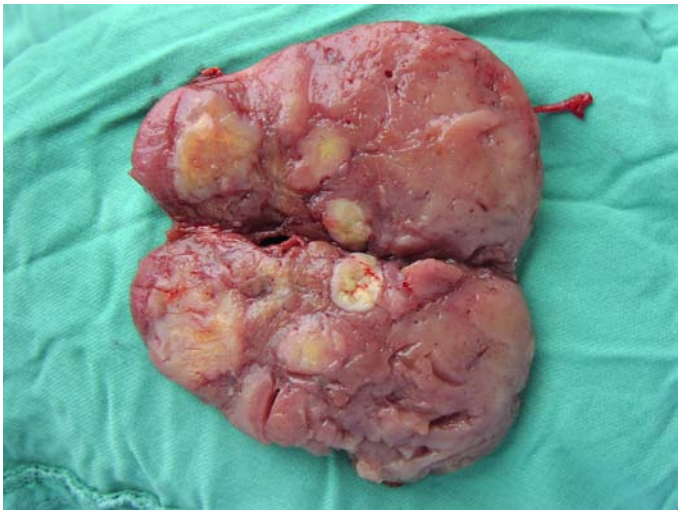

Figure 3: Hemangiopericitoma in mental region (postoperative)

3. Conflict of interest

None

4. Funding

No funding was received

5. Consent

Clinical photos are published with patient consent.

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