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Cerebellar Hemangiopericytoma: A Case Report and Literature Review

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Abstract **Case Report**

Background: Haemangiopericytoma (HPC) is a rare mesenchymal tumour. It was first described in 1942 by A.P. Stout and M. Murray. It represents less than 1% of intracranial tumours. Its location in the cerebellopontine angle and its appearance on CT or MRI can be misleading and lead to a false diagnosis of acoustic neuroma or meningioma. Hemangiopericytoma is characterised by its malignant potential, high rate of recurrence and distant metastases, justifying wide surgical removal and additional radiotherapy.

Keywords: Cerebellar, Hemangiopericytoma, tumour intracranial tumours.

INTRODUCTION

Haemangiopericytoma is a rare mesenchymal tumour arising from Zimmerman's pericytes: contractile cells surrounding the capillaries. It was first described in 1942 by A.P. Stout and M. Murray [1]. It represents less than 1% of intracranial tumours. Its location in the cerebellopontine angle and its appearance on CT or MRI can be misleading and lead to a false diagnosis of acoustic neuroma or meningioma. Hemangiopericytoma is characterised by its malignant potential, high rate of recurrence and distant metastases, justifying wide surgical removal and additional radiotherapy.

CASE REPORT

Mrs. Y.B, 51 years old, without any particular pathological history, presented an intracranial hypertension syndrome, associated to a cerebellar syndrome. A cerebral CT scan was performed showing an extra-axial left cerebellar lesion suggestive of a meningioma, however a metastatic lesion could not be excluded. A cerebral MRI was performed which showed a mass of the cerebellar hemisphere measuring 23*27*40 mm suggestive of a meningioma, having an intimate contact with the homolateral lateral sinus. The patient underwent surgery with a total removal of the tumor with spindle cells. The morphological aspect of the tumor was evocative of a solitary fibrous tumor. On immunohistochemical study, WHO GIII hemangiopericytoma, anti 34 positive AC, anti STAT 6 positive AC, anti EMA negative AC, anti GFAP negative AC, Ki 67 at 10%. Postoperative MRIs and 3 months of follow up showed no evidence of recurrence or residual disease**.**

DISCUSSION

Hemangiopericytomas of the central nervous system are rare: they represent less than 1% of intracranial tumours and 2 to 2.4% of meningeal tumours [2, 3]. Localization in the cerebellopontine angle is very rarely reported in the literature [4, 5]. HPC usually occurs in the fifth decade, but without gender predilection [14]. Intracranial HPC tends to occur at earlier ages than meningiomas [6]. Their distribution is similar to that of meningiomas [7, 8]. They are mainly located in the supratentorial region and are often parasagittal in contact with the venous sinuses.

The clinical symptomatology, which is not characteristic, depends on the location and size of the tumour. CT and MRI are not very specific and cannot categorically distinguish between HPCs and meningiomas given the similarity of their radiological characteristics; however, they provide important information concerning the size of the tumour, its location, its relationship with adjacent structures and the extent of its vascularisation in order to guide their therapeutic action. Angiography is of particular interest because of the vascular origin of haemangiopericytoma. It can sometimes be used for preoperative embolisation, or rarely postoperatively, after surgical resection of the tumour with persistent arterial bleeding [9]. However, the diagnosis of certainty is based on anatomopathological and immunohistochemical study;

the tumour cells are spindle-shaped and show 12q13q inversions and overexpression of the NAB2-STAT6 gene fusion [15].

The treatment of haemangiopericytoma is primarily surgical. Other methods (embolisation, irradiation or chemotherapy) can only be complementary, aiming to improve the local and general prognosis [10,11]. The operative difficulties are essentially related to the hyper vascularisation of the tumour and the causes of the limit of total excision, in particular the location of the tumour and its extension to neighbouring organs. The total resection rate reported in the literature varies between 50% and 83%. However, total resection of the tumour is not synonymous with no recurrence and no metastasis [11]. Operative mortality varies from 0 to 27% and some deaths are secondary to bleeding, which is why some authors recommend preoperative embolisation when the diagnosis of haemagiopericytoma is suspected [10]. Haemangiopericytomas are more radiosensitive than meningiomas. Initially reserved for unresectable or metastatic tumours, radiotherapy is now performed almost systematically in addition to surgery [4]. Numerous studies have shown that post-operative radiotherapy significantly reduces the percentage of recurrences and improves the prognosis of patients [16]. Dufour *et al*. [10] report recurrence rates of 12% in the irradiated group versus 88% in the non-irradiated group. Guthrie *et al*. [13] report recurrence rates of 52% in irradiated patients versus 86% in non-irradiated patients. The local recurrence rate varies from 26% to 80% and depends on the quality of the excision, the duration of follow-up and the use of postoperative radiotherapy. Dufour H et al report an average recurrence rate of 45%. The average recurrence rate was 38.7% for Jeong HoonK *et al*. [12]. The majority of these recurrences occur locally at the initial tumour site and more rarely at a distance [12]. The most common metastatic locations are bone, lung and liver [13]. Overall survival at 5 years varies between 67% and 96%, with a median of 7 to 16.2 years [15]. This requires strict long-term surveillance based on a thorough clinical examination, chest X-ray, abdominal ultrasound or pet scan when indicated [10, 12].

CONCLUSION

HPC is a rare tumour whose behaviour is unpredictable and whose evolution is marked by the occurrence of recurrences, the time of appearance of these recurrences is very variable which means that long-term surveillance is required. The clinical presentation of haemangiopericytoma is non-specific. Imaging (CT and MRI) can be misleading and lead to a false diagnosis of meningioma. Only pathological examination can decide and give the exact diagnosis by histology and immunohistochemistry. Treatment is based on surgery and radiotherapy. These are tumours with a high potential for recurrence and metastasis, requiring prolonged follow-up.

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