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Intracerebral Masson's Tumour: A case report

Veejay Bagga¹, Ahilan Kailaya-Vasen¹, Parag Sayal¹, Stephen B Wharton² & Umang Patel¹

Department of Neurosurgery¹ & Histopathology²
Royal Hallamshire Hospital,
Sheffield
UK.

Corresponding author – Veejay Bagga

Address: Department of Neurosurgery, Royal Hallamshire Hospital, Glossop Road, Sheffield, UK.

Email: vbagga@doctors.org.uk

Abstract

We report a case of a 24 year old male patient with a 3 year history of headache and headache. Neuroradiological imaging showed a slow-growing lesion consistent with a low-grade glioma. Histological analysis following resection revealed intravascular papillary endothelial hyperplasia (Masson's tumour).

The diagnosis of this lesion is difficult as it can mimic a neoplastic lesion. However, once a diagnosis is confirmed, conservative as well as surgical treatment options should be carefully considered. Patients with subtotal resection must undergo long-term follow-up surveillance imaging as recurrence is a possibility.

Introduction

Intravascular papillary endothelial hyperplasia, also known as Masson's tumour, is an unusual, benign vascular lesion which is characterised by reactive proliferation of endothelial cells within an organised thrombus¹. Although relatively uncommon, it is most often seen in skin and soft tissue where it resembles a sarcoma². However, it can arise from any vascular origin in the body and has been described in a variety of locations such as the oral cavity^{3,4}, ulnar artery⁵, internal auditory canal⁶, bladder⁷, hand⁸ and skull⁹. The central nervous system can also be affected by this lesion. Reports have described these lesions causing spinal cord compression¹⁰ and cauda equina syndrome¹¹. Intracranial lesions are however extremely rare, with only 20 intracranial cases having being reported¹². Here we report a further case, describing a 24 year old who presented with a 3 year history of intermittent headaches and dizziness.

Case Report

A 21 year old, left-handed male underwent a CT head and MRI following episodes of intermittent headache and dizziness. Imaging revealed a small enhancing lesion within the left inferior frontal lobe (fig.1). Given the size and location of the lesion, it was felt that this was an incidental finding and an unlikely cause of his headache. He was therefore managed conservatively.

3 years later, he attended his general practitioner complaining of a change in the character and nature of his headache, describing it as being a more constant, left-sided pain which often woke him up at night. In addition, he described some non-specific left-sided upper limb numbness and sluggish eye movements. He was referred to a neurology outpatient clinic for further assessment where clinical examination was normal and fundoscopy showed the appearance of the optic discs to be unremarkable. He was subsequently given the diagnosis of 'chronic daily headache' and was managed conservatively. For completion, a repeat MRI was performed which showed that the lesion which had been identified 3 years earlier had increased in size by 1mm (fig.2). He was therefore discussed in the neuro-oncology MDT for a definite long-term management plan with regards to the intracerebral lesion. A wide differential diagnosis was proposed which included a dysembryoplastic neuroepithelial tumour (DNET) or a low-grade glioma. The patient was subsequently offered both conservative and surgical management options and he chose the latter. He underwent image-guided craniotomy and excision of the lesion. Intra-operative appearance of the lesion represented a globular vascular structure possibly representing a venous varix. Histological analysis revealed an ovoid lesion derived from a single blood vessel, the lumen of which showed papillary structures with hyaline collagenous cores covered by plump endothelial cells, which were immunoreactive with antibodies to CD31. Dystrophic calcification was present. Immunohistochemistry to smooth muscle actin demonstrated smooth muscle in the wall of the circumferential vessel as well as some reactivity within intravascular papillary structures. These appearances are characteristic of intravascular papillary endothelial hyperplasia (Masson's tumour) (fig.3).

Discussion

In 1923, Pierre Masson first described a vascular lesion within an ulcerated haemorrhoidal vein of a 68 year old man and termed it '*Hemangioendotheliome vegetant intravasculaire*'¹³. Over the past 90 years, it has been referred to by a variety of names and is now most commonly referred to as *intravascular papillary endothelial hyperplasia* or *Masson's lesion*. It is characterised by papillary proliferation of the vascular endothelium and can be described as being: a) Primary, if arising from a normal vessel, b) Secondary, if originating from a pre-existing vascular condition e.g. aneurysms^{14,15} or c) if extravascular in nature⁴.

The occurrence of this benign lesion is relatively rare, particularly when associated with the central nervous system. In fact, only 20 intracranial cases have been reported to date¹². Interestingly, this lesion does not show age-related characteristics as individuals of all ages, including neonates, have

been shown to be affected¹⁶⁻¹⁸. However, it does seem to have a slight female predominance^{12,17}.

The diagnosis is often difficult because, as highlighted in this case, radiological appearance is similar to other, more sinister lesions such as gliomas. In fact, a diagnosis can only be confidently made when a tissue specimen is available for histological analysis. As these lesions are benign and typically indolent, conservative management should be considered, especially if there is a high-risk of severe, long-term neurological deficit. Surgical excision however, may be the only option and complete resection is often curative¹⁷, unlike subtotal resection which is associated with recurrence¹⁹. Where complete resection is not possible, adjuvant Gamma knife radiosurgery should be considered as this has been shown to be an effective²⁰. However, one must proceed with caution as radiosurgery has been suggested to be one of the predisposing factors associated with this lesion²¹.

In conclusion, the diagnosis of intracerebral intravascular papillary endothelial hyperplasia is difficult as it can mimic a neoplastic lesion, both clinically and radiologically. However, once a diagnosis is confirmed, conservative as well as surgical treatment options should be carefully considered. If surgery is undertaken, we suggest subtotal resections should be followed with radiosurgical intervention and patients must have long-term follow-up with surveillance imaging as recurrence is a possibility.

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