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Delayed presentation of an arteriovenous malformation after cerebellar hemangioblastoma resection—Case report

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ABSTRACT

INTRODUCTION: Haemangioblastoma has been uncommonly reported to occur in coexistence either temporally or spatially with the development of an arteriovenous malformations (AVM). We present a case of a delayed AVM following haemangioblastoma resection.

PRESENTATION OF CASE: 44 year old female initially presented with a several week history of headaches, vertigo and nausea and emesis and was found to have a cystic lesion with a solid enhancing component on Magnetic Resonance Imaging (MRI) in the superior aspect of the vermis. She underwent gross total resection and final pathology was consistent with WHO grade I haemangioblastoma. One year later, patient re-presented with headaches, dizziness and left trochlear nerve palsy with rotary nystagmus. Imaging revealed a left posterior tentorial paramedian cerebellar vascular nidus with venous drainage into the left transverses sinus suspicious for arteriovenous malformation. She underwent gross total resection of the lesion. Final pathology confirmed the diagnosis of an arteriovenous malformation.

DISCUSSION: Recent research supports both haemangioblastoma and AVM are of embryologic origin but require later genetic alterations to develop into symptomatic lesions. It is unclear in our case if the AVM was present at the time of the initial haemangioblastoma resection or developed de novo after tumor resection. However, given the short time between tumor resection and presentation of AVM, de novo AVM although possible, appears less likely.

CONCLUSION: AVM and haemangioblastoma rarely presents together either temporally or spatially. We present a case of a delayed AVM following haemangioblastoma resection. More research is needed to elucidate the rare intermixture of these lesions.

1. Introduction

Haemangioblastoma has been infrequently reported to occur in coexistence either temporally or spatially with the development of an arteriovenous malformations (AVM) [1]. The first report was in 1965 by Raynor and Kingman where they described a case of coexistence of cerebellar AVM and haemangioblastoma [2]. Since that time a limited number of related neoplasms and vascular malformations have been reported including oligodendroglioma and astrocytoma with AVM and cavernous malformations. Medvedev et al. reported the only other coexistent haemangioblastoma and AVM in the cerebellum in 1991 [1]. We report a case of cerebellar AVM development after resection of cerebellar haemangioblastoma.

2. Case presentation

Patient is a 44-year-old female with past medical history of recurrent sinusitis, malaria, and Hepatitis A and who initially presented to the emergency department with a several week history of headaches, vertigo, nausea and vomiting. On physical examination, the patient was neurologically intact.

A contrast enhanced Computerized Tomography (CT) scan of the head demonstrated a midline posterior fossa cystic lesion with surrounding vasogenic edema resulting in effacement of the fourth ventricle and obstructive hydrocephalus (Fig. 1a). Magnetic resonance imaging (MRI) confirmed a cystic lesion with a solid enhancing $1.6 \times 1.4 \times 1.1$ cm component in the superior aspect of the vermis near the tentorial edge (Fig. 1b–d). She was taken to the operating room for resection the following day. In the sitting position, a stereotactic supracerebellar infratentorial approach was performed for resection of the solid portion of the mass after drainage of the cystic portion. We noted the utmost importance of the close proximity of this tumor to the internal cerebral veins and basal veins of Rosenthal. Post-operative MRI demonstrated gross total resection (Fig. 2). Final pathology confirmed World Health

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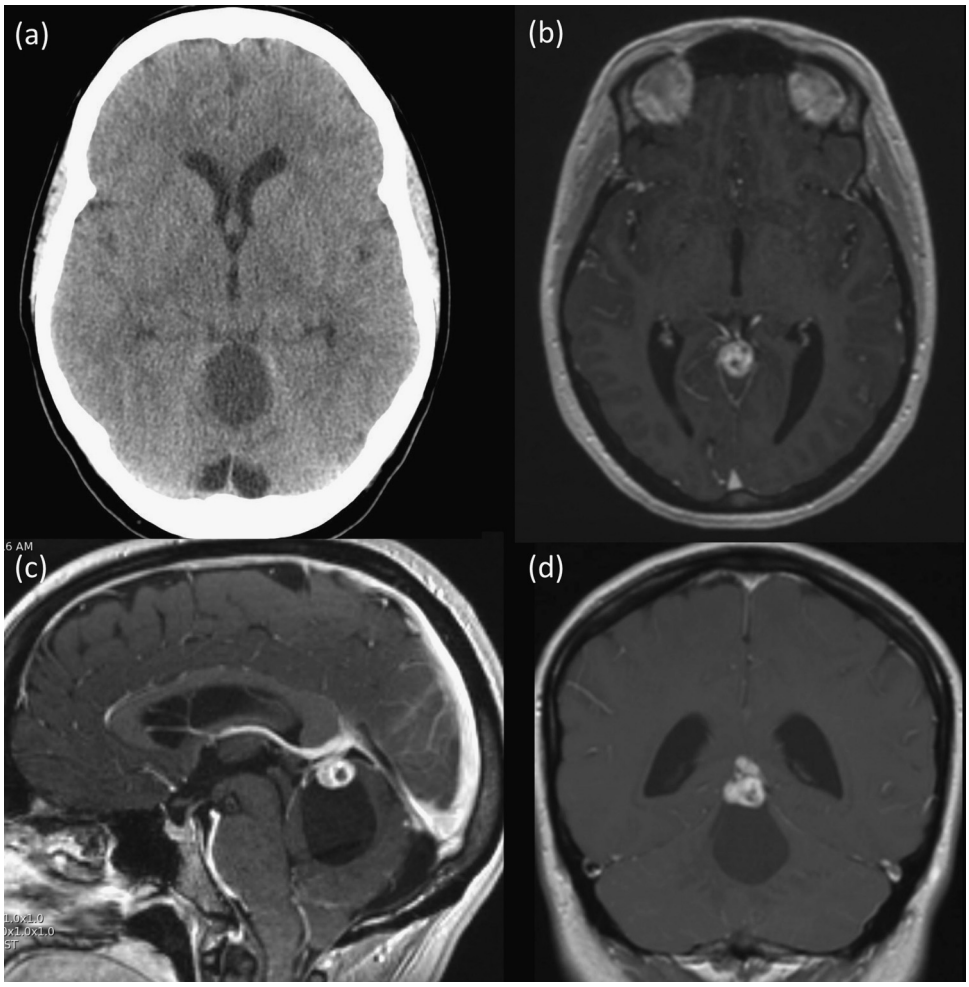

Fig. 1. (a) Axial CT without contrast demonstrating cystic midline posterior fossa lesion with mild obstructive hydrocephalus. Axial (b), sagittal (c), and coronal (d) volumetric T1 with gadolinium MRI demonstrating enhancing lesion at the tentorial apex adjacent to the vein of galen with large cystic component with the cerebellum.

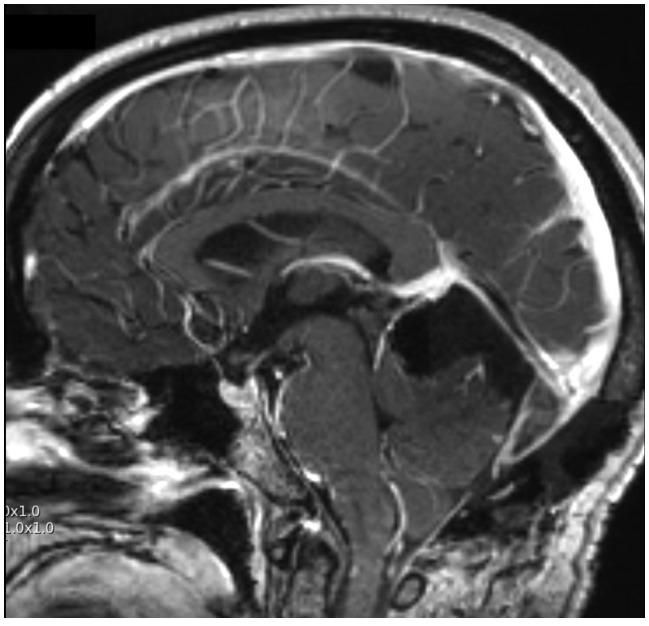

Fig. 2. Postoperative sagittal volumetric T1 with gadolinium demonstrating gross total resection after supracerebellar infratentorial approach.

Organization (WHO) grade I haemangioblastoma. Patient had an unremarkable post-operative course and was discharged home on post-operative day 3. At her 6 week and 6 month post-operative visits, she was doing well. She underwent CT abdomen, dilated ophthalmologic examination, and full neuroaxis MRI to rule out any additional lesions suggestive of von-Hippel-Lindau disorder, all of which were unremarkable. Repeat MRI revealed no recurrence of the lesion (Fig. 3). Of note, she did have intermittent vertigo and headaches following her original resection. She was diagnosed with migraines and they improved with conservative management.

One year after her original surgery, she presented to the emergency room after a 2-day history of headaches with increased dizziness resulting in a fall from standing. Neurological examination showed left trochlear nerve palsy and rotary nystagmus. There were no other neurological deficits. CT scan of her head demonstrated an acute left tentorial subdural hematoma as well as a small left cerebellar intracranial hemorrhage (Fig. 4a). Further CT angiography and MRI demonstrated a nodular focus of enhancement on the medial margin of the left cerebellar hemorrhage raising concern for new or recurrent haemangioblastoma (Fig. 4b–d). Diagnostic digital subtraction angiogram (DSA) was then performed and demonstrated a $2.4 \times 0.9 \times 1.0$ cm left posterior tentorial paramedian cerebellar vascular nidus primarily supplied by bilateral posterior inferior cerebellar arteries and left superior cerebellar artery with venous drainage into the left transverses sinus suspicious for arteriovenous malformation (Fig. 5a, b). In addition, there was a 2.5×2.0 mm intranidal aneurysm (Fig. 5c, d). There-