

## Bilateral acquired ptosis, an unusual presentation of a cerebral arteriovenous malformation

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### Abstract

Arteriovenous malformations (AVM) of the brain are uncommon congenital lesions with a complex array of arterial to venous shunts. The common clinical manifestations include haemorrhage, seizures, headache, progressive neurologic deficits and congestive cardiac failure. We report a child who presented with bilateral acquired ptosis, a very rare presentation of AVM which has not been previously reported.

### INTRODUCTION

Arteriovenous malformations (AVM) of the brain are uncommon congenital lesions with a complex array of arterial venous shunts. They vary in size, but are often large, involving the cortex and white matter. The common clinical manifestations include hemorrhage, seizures, headache, congestive heart failure and progressive neurological deficits depending on the location of the malformation.<sup>1</sup> We report a child who presented with bilateral acquired ptosis, an unusual manifestation of AVM. The diagnosis was made by magnetic resonance imaging (MRI) and confirmed by digital subtraction angiography (DSA).

### CASE REPORT

This 8 years old child was referred for evaluation of difficulty in walking for 2 ½ years.. He was the third child of a nonconsanguineous marriage. Antenatal and postnatal periods were uneventful. His parents noticed that since the age of 5 ½ years, he has motor difficulty becoming worse towards evening. At age of 6 years, he would trip over objects and would frequently fall. He had difficulty keeping up with his friends when playing. Running, jumping, and hopping were also abnormal. His gross motor development at this stage was normal. The parents also gave history of drooping of both eyelids, symmetrical in onset, severe, partial and did not vary during the day (Figure 1). The ptosis was not present at birth, it only developed later in life. On examination, he had bilateral ptosis and poor fine motor coordination. There was levator palpebrae

superioris weakness both sides. The cover test for both eyes were normal showing no extra ocular muscle weakness. Pupils were normal in size, reacting to light. There was no light-near dissociation. There was no nystagmus or papilledema or muscle weakness. The rest of the neurological examination including cerebellar sign was unremarkable. Auscultation did not reveal any bruit. A diagnosis of possible myasthenia gravis was made. Acetylcholine receptor antibodies titer was normal, the neostigmine test was negative, and electromyography including repetitive stimulation test and nerve conduction velocities were all normal. MRI of the brain showed a large complex AVM with multiple vessels in the thalami, brainstem and cerebellum (Figures 2-5) which was confirmed by the DSA. The vein of Galen was markedly dilated and was probably the venous channel of the AVM. The arterial supply appeared to be from the posterior vertebrobasilar circulation as both of the posterior communicating vessels were enlarged. The child was referred to neurosurgery service, but was advised against surgical treatment. Endovascular therapy was also thought to be unlikely to benefit patient. The ptosis did not improve during follow-up.

### DISCUSSION

AVMs are an admixture of arteries and veins. They can range from few centimeters to few millimeters in size. Malformations result from failure of the formation of the capillary bed between primitive arteries and veins in the brain during the first trimester of fetal life. The incidence of AVM in children is 1 in 100,000 and approximately 10%

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Figure 1. Picture of patient showing bilateral severe ptosis.

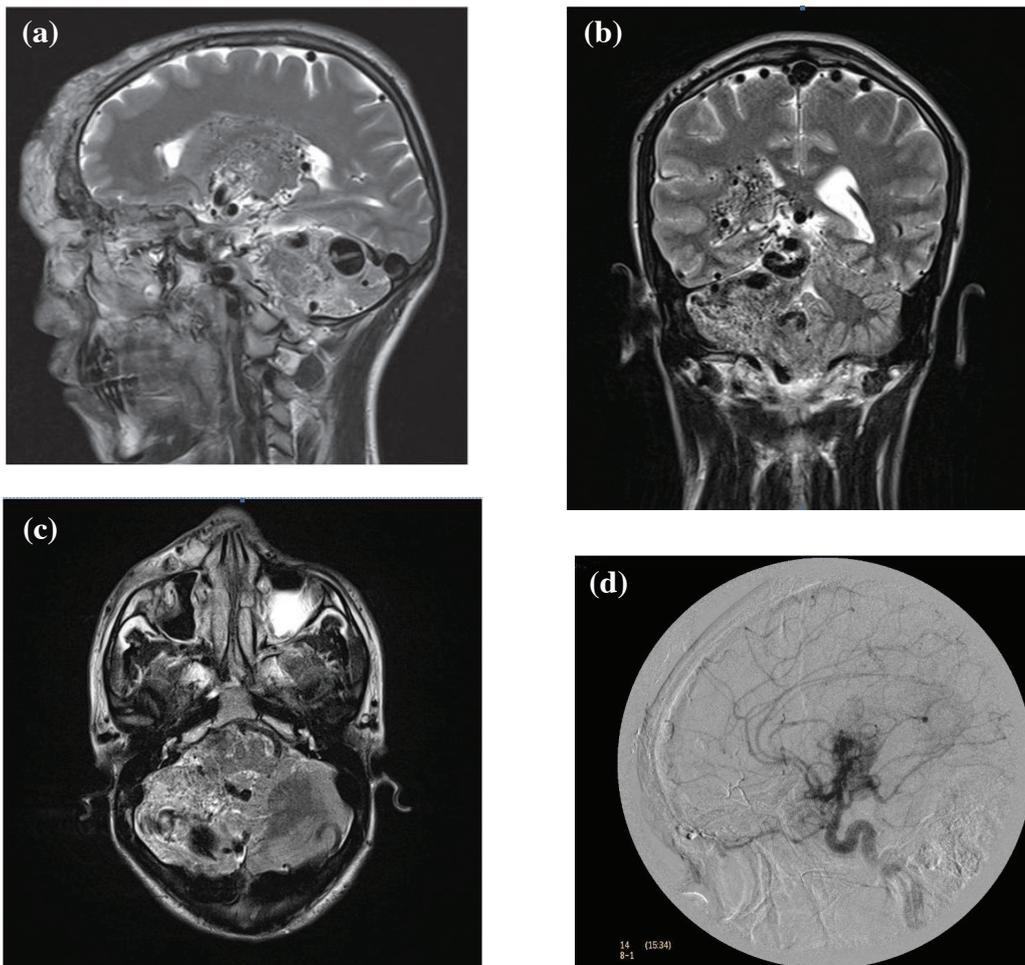


Figure 2. MRI brain in (a) sagittal, (b) coronal, (c) axial view, and (d) DSA showing a large complex AVM.

to 20% become symptomatic during childhood.<sup>2</sup> Hemorrhage occurs in 74% and a chronic seizure disorder reported in 12% of patients in a 40-year study of 132 patients.<sup>3</sup> Other features depend on the location of malformation.

Ptosis during presentation of AVM is rarely seen<sup>4</sup>, the exact incidence is not known. Moody and Poppen<sup>5</sup> mentioned visual deficits in 30 of their 105 cases with AVM, but no cases with bilateral ptosis were mentioned. Many of the earlier reports spoke of chronic unilateral ptosis, papilledema and visual loss.<sup>5-8</sup> However, bilateral ptosis at presentation has not been reported in the literature.

The MRI of our patient showed evidence of a large complex brain AVM with multiple vessels in the thalami, brainstem and cerebellum. The vein of Galen was markedly dilated and was probably the venous channel of AVM. The bilateral ptosis is most likely due to siphoning of blood flow away from adjacent brain tissue (the “steal phenomenon”), affecting the dorsal midbrain. The other possibility is compression of the dorsal midbrain that is responsible for levator palpebrae superioris function. However, dorsal midbrain dysfunction could produce light-near dissociation and up gaze paresis, these were absent in our patient. Bilateral ptosis due to dilatation of posterior communicating vessels causing third nerve injury is another possibility.

In conclusion, AVMs can present with a wide spectrum of manifestations ranging from subtle features to devastating neurologic symptoms. This case highlights the importance of doing an early MRI scan for children presenting with unexplained bilateral acquired ptosis.

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