



Cerebellar mutism after embolization of vermian arteriovenous malformation

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Abstract

Background Cerebellar mutism is usually associated with posterior fossa tumor surgery.

Case Report We report a case of a 17-year-old female, presented with headache and tremor after hemorrhage from a vermian arteriovenous malformation. She was successfully treated by embolization; however, on immediate postoperative, she developed persistent mutism. To the best authors' knowledge, this is the first case of cerebellar mutism after endovascular treatment reported in the medical literature.

Conclusion The endovascular approach may have the same potential of complication of conventional surgery; therefore, more study is necessary to clarify the role and limits of this technique to treat cerebellar arteriovenous malformation.

Keywords Mutism · Intracranial arteriovenous malformations · Endovascular procedures · Postoperative complication · Therapeutic embolization · Intracranial hemorrhage

Introduction

The term cerebellar mutism (CM) is used to describe the muteness after cerebellar injury, in opposition to lesions in the cerebrum or the lower cranial nerves [8]. Although CM is usually associated to “open” surgery for posterior fossa tumors in children, it was already described after trauma, infections, and cerebellar hemorrhage, either from arteriovenous malformation (AVM) or tumor [1, 3, 5–7]. Therefore, it is possible that the endovascular approach has the same potential of developing such complication. We report a case of postoperative CM after embolization of a vermian AVM.

Case report

A 17-year-old female patient complained of sudden onset of severe headache and evolved with impaired consciousness. At admission, she presented with somnolence, confusion, Glasgow coma scale 13, and distal limb tremor. Investigation with CT scan and MR showed posterior fossa hemorrhage in the vermis and 4th ventricle (Fig. 1). The patient underwent cerebral angiography that revealed an AVM located at the cerebellar culmen, mainly fed by the superior cerebellar artery. At the same surgical time, under general anesthesia and full anticoagulation, a

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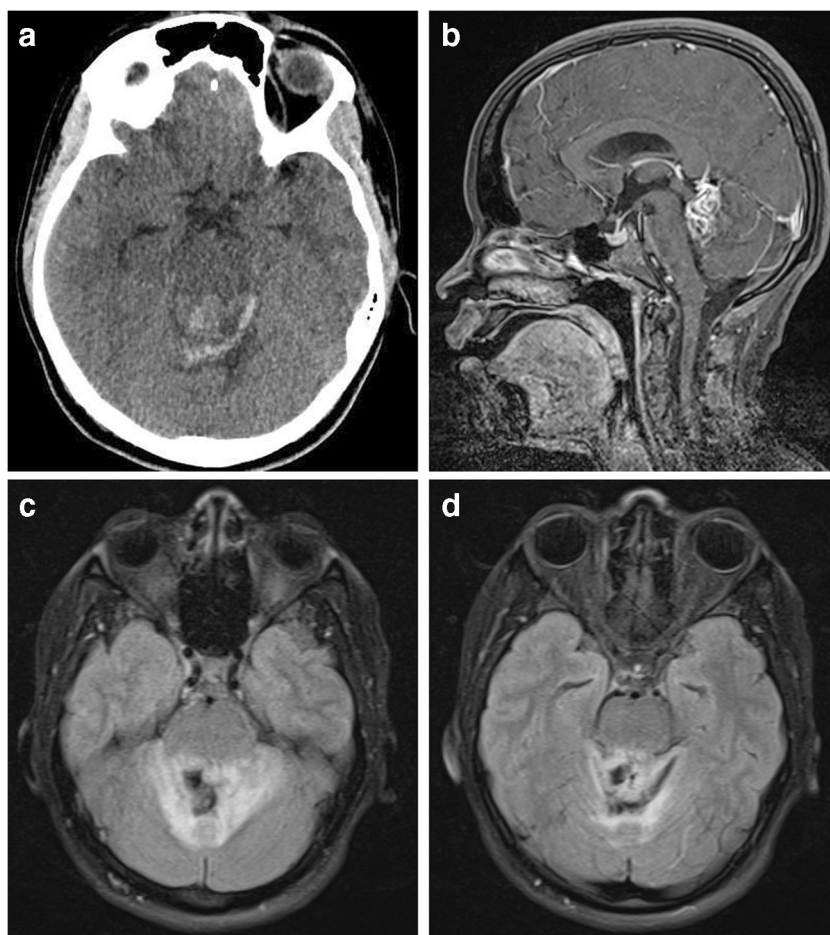
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Fig. 1 Preoperative imaging shows cerebellar bleeding in the vermis and 4th ventricle on CT scan (**a**); flow voids and hemorrhage with circumjacent edema on MR post-Gadolinium T1 (**b**) and FLAIR (**c, d**)



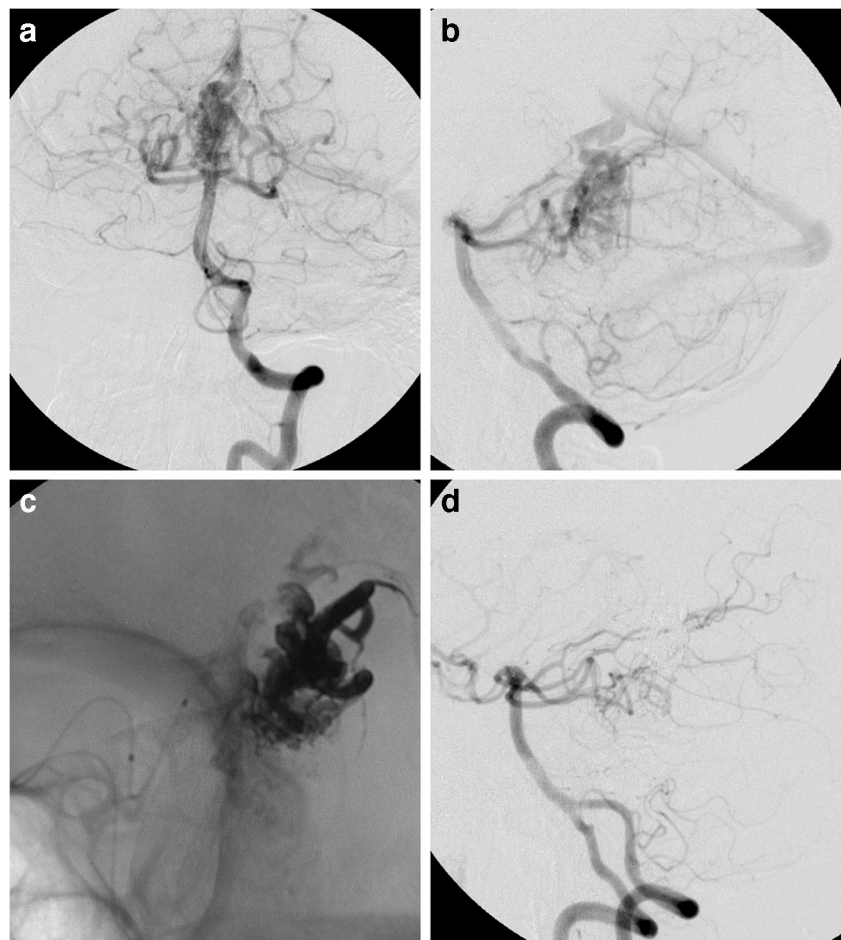
guiding catheter (Neuron™ 6F, Penumbra Inc., Alameda, CA, USA) was positioned into the distal segment of the left vertebral artery; a microcatheter (Apollo™ 10, Medtronic, Irvine, CA, USA) over a microguidewire (Silverspeed™ 0,010, Medtronic) was placed into the nidus; embolization was performed using non-adhesive EVOH-based liquid embolic agent (Squid-12™, Emboflu, Gland, Switzerland) achieving total occlusion (Fig. 2). Despite satisfactory angiographic result, the patient evolved postoperatively with mutism and right tremor. On control CT scan and MR, no ischemic or hemorrhagic changes were noted (Fig. 3a); thus, she was discharged on the 10th postoperative day. At 6-month follow-up, the patient preserved understanding and cognitive functions; however, she still presented mutism, dysmetria, dysidiadochokinesia, and tremor of head and limbs. She was able to stand up and walk with support, although with gait ataxia. Control MR showed gliosis and encephalomalacia adjacent to the arteriovenous malformation, evolving portions of the pons, rhomboid fossa, superior cerebellar peduncles, and cerebellar vermis (Fig. 3b–d). The tractography was inconclusive.

Discussion

The terms CM, pseudobulbar palsy, posterior fossa syndrome (PFS), and cerebellar cognitive affective syndrome (CCAS) are used to designate the complex neurological condition usually following surgery of cerebellar and fourth ventricular tumors [11]. Although these terms may be sometimes interchangeable, since clinical presentation depends on anatomical structures and neural circuits, it is consensus that designation PFS lacks precision and specificity and should be retired [11]. The CM is not a prerequisite to develop CCAS, suggesting involvement of different circuits; therefore, the circuit disruption associated with CM needs to be distinguished from that causing CCAS [11].

The dentato-thalamo-cortical (DTC) tract is an important outflow pathway from the cerebellar nuclei towards the cerebral cortex, and its interruption is accepted as the main cause for CM [8]. The DTC tract extends from the dentate nucleus via the contralateral red nucleus and thalamus to reach the contralateral cerebral cortex [8]. The proximal portion of the DTC pathway, known as the proximal efferent cerebellar pathway (pECP), has been implicated in the development of CM,

Fig. 2 Digital angiogram shows a midline arteriovenous malformation filled by superior cerebellar arteries (**a, b**); the nidus was fulfilled with liquid embolic agent (**c**); achieving total occlusion of the lesion (**d**)



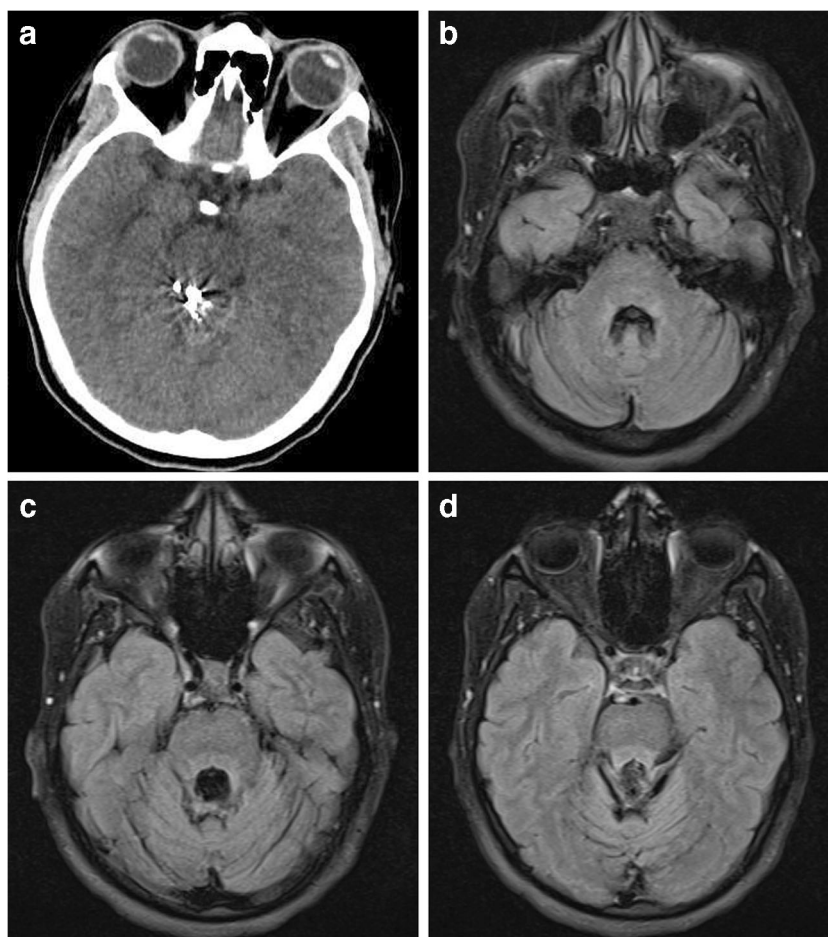
and it is constituted by the dentate nucleus, superior cerebellar peduncle (SCP), and mesencephalic tegmentum [8]. In our patient, the upper vermis was involved, whereas CM was already associated with injuries to the middle cerebellar peduncle (MCP), brachium pontis, dentate nucleus, and inferior, posterior, and upper vermis [2, 3, 7, 10]. The exact pathogenesis of CM remains unknown, with evidence pointing to a multifactorial disorder [2]. Surgical manipulation causing direct injury to anatomical structures, especially the pECP, has been associated to CM [2]. In the last years, however, some studies have questioned the sole role of surgery in the pathogenesis of CM [4], and case reports such as the present one may further confirm this hypothesis and suggest the need for a deeper understanding of CM pathogenesis. As CM occurred without surgery, it was first attributed to an ischemic injury secondary to endovascular treatment affecting the DTC tract, since the caudate nuclei were preserved. Instead, this could hypothetically be also a consequence of the initial hemorrhage, as already reported [1, 3, 7], even presenting delayed onset. The alternative relation with vasospasm has not been considered on the literature.

Since most of the cerebellum is not related to motor control, the deep cerebellar nuclei are involved in limbic and

autonomic circuits (fastigial nucleus) as well as cognitive domains (ventral part of the dentate nucleus) [11]. Emotional lability is a frequent neuropsychiatric feature; however, impairments may occur in the modulation of five domains of behavior: attention, mood, social skills, psychosis, and autism spectrum disorders [11].

Immediate postoperative MR may show parenchymal edema along the surgical margins, characterized by increased T2 signal; DWI can distinguish cytotoxic edema due to cellular swelling, with restricted diffusion from vasogenic edema, due to an increase in extracellular fluid, with unrestricted diffusion [2]. Careful clinical observation may be correlated with the brain imaging to perform detailed structure function analyses to identify and understand the neural circuits, prevent complication, and develop new methods of treatment [11]. Midline location and brainstem invasion on preoperative imaging are consistent identifiable risk factors of CM [2]. Furthermore, five significant risk factors including the fourth ventricular tumor location, bilateral MCP invasion and/or compression, dentate nucleus invasion, and age >12.4 years were considered to develop an assessment scoring system and predict the risk CM on

Fig. 3 The 6-month postoperative imaging: CT scan shows reabsorption of the hemorrhage and embolic material (a); the MR FLAIR demonstrates small gliosis in the pons, cerebellum, and superior cerebellar peduncles, adjacent to the arteriovenous malformation (b–d)



preoperative MR, but they need to be validated [9]. On postoperative imaging, bilateral pECP injury was identified as a significant risk factor to develop CM [2]. Our patient presented age >12.4 years, midline location AVM, and bleeding into the 4th ventricle as possible risk factors; however, the complication was not anticipated since no case had been reported to the better of our knowledge.

Considering the absence of an effective treatment established to improve CM recovery, it is primarily based on clinical support measures [8]. Although pharmacological intervention does not achieve desired effect in all cases of CM, bromocriptine may be helpful in some patients, considering the response of reversing symptoms of akinetic mutism [8]. There are few language therapies reported in the literature; however, speech rate is used for evaluation and monitoring the manifestations of CM [8].

Conclusion

CM may complicate the clinical history of posterior fossa AVM, as a consequence of hemorrhage or as a complication

of embolization. Accordingly, physicians should keep in mind this complication and parents should be adequately informed. Further understanding of pathogenetic mechanisms and underlying anatomical circuit of CM is required to prevent and treat this complication.

Author contributions All authors contributed to the study conception and design. Dr. Schmitz and Dr. Ribas performed material preparation, data collection, and analysis. Dr. Chula prepared the images and achieved the informed consent, while Dr. Demartini wrote the first draft of the manuscript. Dr. Gatto and Dr. Koppe made critical revision of the manuscript. All authors commented on previous versions of the manuscript and also read and approved the final manuscript.

Compliance with ethical standards

Conflict of interest The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Informed consent A parental permission in research with children was achieved, and the informed consent was sent attached.

Research involving human participants The study was performed in accordance with the ethical standards; all patient identification was removed to preserve anonymity.

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