Posterior Fossa Syndrome Secondary to Ruptured Cerebellar Arteriovenous Malformation: A Case Report

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Abstract
Posterior Fossa Syndrome (PFS) is well known surgical complication of posterior fossa pathologies, mainly in pediatrics group. We report 10 years old girl who presented with sudden onset of headache, vomiting and decrease in level of consciousness. Upon arrival to emergency department, her GCS was 7/15. Patient was resuscitated and intubated. Initial brain CT revealed IVH with large volume cerebellar hemorrhage on the left lateral hemisphere extending to the vermis and associated hydrocephalus. CT angiography showed large cerebellar AVM. Patient was admitted to PICU and an external ventricular drainage was inserted. Few days later, she underwent cerebral angiography and embolization of AVM with 80% obliteration rate. After extubation, patient showed symptoms and signs of PFS that lasted six weeks before she returned gradually to her normal status. To our best knowledge, this is the second case with ruptured cerebellar AVM that was followed by PFS. The exact mechanism remains unclear.

Introduction
Cerebellar mutism or posterior fossa syndrome secondary to arteriovenous malformation is extremely rare [1,2]. In the literatures, there are many well known cases of cerebellar mutism or Posterior fossa syndrome (PFS) secondary to posterior fossa tumor resections [3-7], more in pediatrics patients [6]. PFS manifests by diminished speech ability with a progression to mutism, hypotonia, emotional disturbance and ataxia [5]. Tumor that is more localized to midline in the posterior fossa, mainly in the vermis with extension to brainstem is of a significant risk factor [8]. Medulloblastomas, astrocytomas and ependymomas represent the most common types of tumors. The main age in one study was 10.4 years [8-10].

This syndrome is a transient, usually starts from 0 to 6 days post operation and it may continue to up to four months before gradual resolving [9,10]. The recovery range and period is widely variable. Ataxi after 1-2 months post surgery is around 100-85% while hypotonia and third nerve palsy were 50% and 40% respectively [5,11-13]. Motor speech deficits 1 year after the surgery were around 68% [14]. Other reported complications include variable degree of behavioral and cognitive symptoms like depression, antisocial behavior, anxiety, academic difficulties and poor intellectual ability [15-18].

The Case
10 years old girl presented with sudden onset of headache and vomiting that was followed by decrease in level of consciousness. Upon arrival to emergency department at our hospital, her GCS was 7/15. No significant past medical history. She was resuscitated and intubated. Initial brain CT revealed IVH with large volume cerebellar hemorrhage on the left lateral hemisphere extending to the vermis and associated hydrocephalus (Figure 1). CT angiography showed large cerebellar AVM. Patient was admitted to PICU and an external ventricular drainage was inserted. Few days later, she underwent cerebral angiography and embolization of AVM with 80% obliteration rate. After extubation, patient showed signs and symptoms of PFS that lasted for six weeks before she returned gradually to her normal status.
Posterior fossa syndrome (PFS) in this case followed the regular course of those underwent surgical resection of posterior fossa tumor. Ischemia or infarction due to the primary insult or the intervention might be a possible cause for this phenomenon. In our case, we understand that the lesion itself (AVM) is the cause, the real mechanism remain unknown regardless of suggested hypothesis. It’s crucial that (PFS) to be involved in the informed consent for any such pathology involving cerebellum. Treating team, nursing staff and the family should be aware about the possibility of having PFS in their candidate patient for surgical or endovascular intervention involving the posterior fossa. A huge advance has been made in both, the techniques and tools to diagnose and treat neurosurgical patients in the past decade like the introduction of functional MRI. Using tractography technique may help in future to define the definitive cause and mechanism of PFS and to plan surgical intervention that minimizes the risk of developing (PFS). Pharmacological treatment, speech therapy and weather the involvement of the brain stem by the primary insult, surgical or endovascular intervention remain an area for a further research.

Conclusion

To our best knowledge, this is the third case worldwide with ruptured cerebellar AVM that was followed by PFS. We think it is something to mention to the family and prepare them for such before intervention; however the exact mechanism remains unclear.

References
