Thoracolumbar spinal vascular malformation as a rare cause of isolated intraventricular hemorrhage

Case report

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Spinal vascular malformations are rare vascular lesions that most frequently present with back pain, radiculopathy, and/or myelopathy. Neurological decline is typically secondary to progressive radiculopathy, myelopathy, venous thrombosis, and stroke. Rarely these vascular lesions can present as Foix-Alajouanine syndrome (subacute necrotic myelopathy) or acute intramedullary hemorrhage with secondary subarachnoid hemorrhage (SAH), arachnoiditis, and ischemia.5 Few case reports have described thoracolumbar spinal vascular malformations that present with both subarachnoid and intraventricular hemorrhage. This is the first reported case of a thoracolumbar spinal vascular malformation presenting with isolated intraventricular hemorrhage on initial imaging followed by acute and fatal rehemorrhage.

KEY WORDS
• intraventricular hemorrhage
• spinal arteriovenous malformation
• subarachnoid hemorrhage
• rehemorrhage
• hydrocephalus
• spine
• vascular disorders

Spinal vascular malformations (SVMs) are rare vascular lesions that most frequently present with back pain, radiculopathy, and/or myelopathy. Neurological decline is typically secondary to progressive radiculopathy, myelopathy, venous thrombosis, and stroke.5 Rarely these vascular lesions can present as Foix-Alajouanine syndrome (subacute necrotic myelopathy) or acute intramedullary hemorrhage with secondary subarachnoid hemorrhage (SAH), arachnoiditis, and ischemia.6 A few case reports have described thoracolumbar spinal vascular malformations that present with both SAH and intraventricular hemorrhage (IVH).2,4,9 These cases have been in children and young adults. This is the first reported case of a thoracolumbar vascular malformation presenting with isolated IVH on initial imaging followed by acute and fatal rehemorrhage.

Case Report

History. This 2-year-old previously healthy girl was transferred to our hospital and presented with 4 days of sleepiness, decreased oral intake, emesis that resolved 1 day prior, and diarrhea. She also had 3 days of meningismus manifesting itself as her holding her neck in extension. Since onset, the symptoms had been stable without worsening. She was afebrile and an infection workup at the initial hospital was negative. The workup did not include a lumbar puncture. There was no history or signs of trauma. There was also no history of back pain, radiculopathy, or myelopathy. Routine coagulopathy workup was normal.

Examination. The patient was awake spontaneously but closed her eyes volitionally when she saw medical personnel. When asleep, she was easily arousable to light tactile stimulation. She was speaking appropriately to her parents and was very purposeful. Her neck was in tonic extension. Her neurological examination was nonfocal. There were no signs of radiculopathy or myelopathy.

Imaging. A head CT scan obtained at presentation to our institution showed a small amount of IVH in both lateral ventricles, the third ventricle, and the fourth ventricle (Fig. 1). The ventricles were mildly enlarged. There was no SAH seen. No source of the hemorrhage was identified on CT, MRI, MR angiography, MR venography, and catheter angiography of the brain and cervical spine. The catheter angiography studies included injections of the common carotid arteries, internal carotid arteries, and vertebral arteries bilaterally.

Hospital Course. Over the next 6 days, while the aforementioned imaging workup was being completed, the patient was observed closely in the ICU and slowly returned to her intact baseline. Although it would be dif-
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Fig. 1. Axial noncontrast CT images obtained at presentation to our institution, showing a small amount of IVH in both lateral ventricles, the third ventricle, and the fourth ventricle. The ventricles are mildly enlarged. There is no evidence of SAH.

Fig. 2. Sagittal (A) and axial (B and C) noncontrast T2-weighted MR images revealing a large thoracolumbar vascular malformation enveloping the spinal cord.

Fig. 3. Axial noncontrast CT images obtained emergently after the second clinical ictus, revealing new extensive cranial and cervical SAH with ventricular dilation.

It was difficult to explain IVH without apparent SAH caused by a thoracolumbar spine lesion, we decided to complete the imaging workup with MRI of the entire spine. This study showed a large thoracolumbar vascular malformation, the likely hemorrhage source (Fig. 2).

After the general anesthesia required for the MRI, the patient was extubated and at her baseline function. Shortly thereafter, she suddenly became unresponsive, showed extensor posturing in all four extremities without twitching, was arching her back, and developed fixed and dilated pupils bilaterally. There was no movement to deep pain. She was immediately reintubated. An emergency head CT scan showed new extensive cranial and cervical SAH with ventricular dilation, presumably from rerupture of the just-diagnosed thoracolumbar vascular malformation (Fig. 3). An emergency ventriculostomy was placed without delay. Just after placement, her examination showed pinpoint pupils bilaterally. She began to move her head slightly. At times, she would breathe spontaneously. She had no corneal reflex, gag reflex, or movement to deep pain.

Over the succeeding 2 days, the ventriculostomy kept her intracranial pressure within the normal range. Her neurological examination, however, did not improve. She was intubated, not sedated, was unresponsive, her eyes were closed; her pupils were equal, round, and reactive to light; she had no corneal reflex, no gag reflex, and no movement to deep central or peripheral pain. She slowly deteriorated from a global hemodynamic perspective, experiencing one ventricular fibrillation and two pulseless electrical activity cardiac arrests from which she was resuscitated. After the third arrest, the parents opted for limitations on future resuscitative attempts. Brain MRI revealed widespread bilateral infarcts of the cerebrum, basal ganglia, thalami, hippocampi, and cerebellum consistent with diffuse global hypoxic-ischemic injury. There were no apparent infarcts in the brainstem. Although one could not have been certain given the three cardiac ar-
rests, the unchanged neurological examination suggested that the infarcts were sustained at the time of the SVM rerupture. Given the grave neurological prognosis, the parents opted to allow natural death and the child died.

Pathological Findings. Postmortem examination revealed diffuse spinal and cerebral SAH secondary to rupture of a large intramedullary and extramedullary vascular malformation. The malformation consisted of an extensive collection of tortuous vessels involving the leptomeninges of the lower thoracic and upper lumbar spinal cord (Fig. 4). An aneurysmal dilation of a lesional vessel was seen (Fig. 4). Microscopic examination identified many vessels from capillary size to larger artery-like and vein-like vessels as well as capillary-sized vessels outside the cord parenchyma (Fig. 5A and B). Similar vessels were seen intertwined within spinal cord parenchyma with no capsule separating the vessels from the cord parenchyma (Fig. 5C–F). A definitive site of rupture of these numerous abnormal vessels could not be identified. However, no other putative source of hemorrhage was found on detailed examination of the CNS. No other systemic vascular lesions were found. Genetic testing for hereditary hemorrhagic telangiectasia was negative.

Discussion

Spinal vascular malformations are rare and represent approximately 10% of CNS vascular malformations in all age groups and 4% of all intraspinal masses. These lesions are directly supplied by radicular arteries and drained by spinal cord veins, although dural supply can occur as with dural arteriovenous fistulas. They are subclassified based on intradural versus extradural location, presence of a nidus, and extramedullary versus intramedullary position. Intramedullary SVMs are analogous to intracranial arteriovenous malformations and may have a feeding artery or intranidal aneurysms predisposing them to rupture.

In contrast to SVMs, spinal cord arteriovenous fistulas (SAVF)s may be intradural or extradural, but they are perimedullary in location. Intradural dorsal SAVFs are more common in adults and typically present with symptoms consistent with venous hypertension; they rarely hemorrhage. Intradural ventral SAVFs may have one or many arterial feeders. These lesions may present with venous congestion and/or hemorrhage. In general, SAVFs comprise about 20% of SVMs and are more common in the pediatric population and in patients with hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu syndrome).

The natural history of SVMs is characterized by venous congestion causing progressive neurological deficits in a majority of patients. Typically these patients present with back pain and sensory or motor changes over months to years. In rare cases, patients younger than 30 years of age can present with sudden myelopathy secondary to hemorrhage. Coup de poignard of Michon refers to sudden back pain due to rupture of an SVM, akin to the thunderclap headache of aneurysmal SAH. Aminoff investigated the risk of rebleeding in SVM patients. Among the 41 patients in their study, 22 (54%) had a second SAH and 4 of these 22 (18%, or 9% of the 41) died. Second hemorrhages occurred as early as within 24 hours after the first episode to well over 5 years subsequently. In other series, investigation of the incidence of hemorrhage of cervical SVMs revealed an even less fa-
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Vorable prognosis: hemorrhage occurred in 78%,1 58%,16 and 57%13 of patients. In the pediatric literature, a thoracolumbar SVM presenting with both SAH and IVH appears to be a rare occurrence.2 There have been case reports of SVMs presenting in the adult population with severe brainstem compression only a short interval.12,18,14 Only 3 case reports of SVMs present in the adult population with severe brainstem compression. In all cases, the clinical, radiographic, and surgical findings suggested that the SVM was the source of the hemorrhage.2,13 In all of these cases there was acute neurological deterioration, but none was caused by rehemorrhage.

Hemorrhagic tumors of the spine can also cause SAH, but this is rare as well. A 2008 review of SAHs from spinal ependymomas noted 18 cases in the literature since 1958.15 Patient age ranged from 14 to 74 years. All but one of these tumors were in the lumbar spine, and all but one presented with signs or symptoms referable to the spine in addition to the cranium. Of the 8 patients who underwent craniotomy, only one had a positive finding. Most of the diagnoses of SAH were based on the results of lumbar puncture. Of the 16 cases, which we will report, the mean was 118 days, the minimum was 5 days, and the maximum was 63 months.

To the best of our knowledge, this is the first case report of a patient in whom initial imaging demonstrated isolated IVH without SAH secondary to a ruptured low thoracolumbar SVM. We do acknowledge that the initial imaging was performed on transfer of the patient to our institution, which was 4 days after the clinical ictus. It is possible that there was SAH whose signal on CT cleared over the intervening days. Nevertheless, the presentation time course and initial prehospital diagnostic difficulty may not be atypical, and therefore apparent isolated IVH may be seen in cases such as this in the future. Recognition of the possibility of a caudal spinal source may allow earlier diagnosis and treatment before catastrophic rehemorrhage.

Although a thoracolumbar SVM is low on the differential diagnosis for isolated spontaneous IVH or SAH, this case suggests that MRI of the entire spine should be considered if cranial imaging does not reveal a source. Especially in children with a smaller distance between the low thoracolumbar and intracranial spaces, cervical spine MRI may not be sufficient. Furthermore, this case demonstrates that these lesions can rehemorrhage after only a short interval.

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Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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