Subarachnoid haemorrhage from Spinal Dural Arteriovenous Fistula in a child

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Background and Objectives:
SAH in the paediatric population is an uncommon entity accounting for 1-2% of SAH in all age groups & only 18% of ICH in the paediatric population. Trauma is the most common cause of isolated SAH in the paediatric population, with ruptured intracerebral aneurysms although still rare, being the most common cause of spontaneous SAH in children. AVMs are uncommon source of spontaneous SAH. Although they are 10 times more common than cerebral aneurysms in the paediatric population, they rarely present with isolated SAH. This report aims to present such a rare case presented to a tertiary care hospital in Trinidad.

Case Presentation:
We present a 3-year-old girl admitted with sudden onset of headaches, weakness of the right leg, photophobia and neck stiffness. Examination revealed MRC grade 2/5 power in the right leg. Non contrast CT scan revealed blood in the subarachnoid space and occipital horns of the lateral ventricles. Cerebral CT angiogram demonstrated neither intracerebral aneurysms nor AVM, but CT angiogram of the spine revealed a left sided DAVF with the feeding vessel entering the spinal canal at the left C6/7 foramen. A C6 and C7 laminoplasty was performed with ligation of the feeding artery. Postoperative obliteration of the DAVF was confirmed radiologically and clinical resolution of leg weakness was noted. The patient developed non communicating hydrocephalus, necessitating the need for a VP shunt.

Conclusion:
Spontaneous subarachnoid haemorrhage in the paediatric population is an uncommon entity with several possible causes. Clinicians must have a high index of suspicion of an underlying lesion.

Introduction
Subarachnoid haemorrhage refers to the extravasation of blood into the subarachnoid space between the pial and arachnoid membranes. It can occur in various clinical grades ranging from low grade with no neurological symptoms to high grade in which the clinical course can be devastating.

In approximately 15% of patients with spontaneous intracranial subarachnoid haemorrhage (SAH), the cause of the haemorrhage is non aneurysmal in origin. Two-thirds of these patients, thus comprising 10% of all patients with intracranial SAH, have a non-aneurysmal perimesencephalic haemorrhage. The remaining 5% of intracranial SAH are caused by a variety of rare conditions which include cerebral arteriovenous malformation.
and dural arteriovenous fistula [1, 2]. Spinal arteriovenous fistulae, in particular if localized in the cervical region, have been reported to present with symptoms and signs suggesting an intracranial cause of the spontaneous SAH [1, 2].

Subarachnoid haemorrhage in the paediatric population is uncommon, accounting for 1% to 2% of SAH in patients of all age groups and only 18% of intracranial haemorrhage in the paediatric population [3, 4, 5]. In the older paediatric population, the symptoms at presentation after a spontaneous haemorrhage are similar to those in adults, including the sudden onset of severe headache (61%), nausea and vomiting (45%), decreased level of consciousness (42%), seizures (26%), and focal neurological deficits (13%) [4]. Younger patients tend to present with increasing irritability and lethargy [6]. Meyer-Heim et al. in a review of paediatric SAH in 2003, noted that the onset of symptoms was acute in 53% of patients and subacute in 47% [4].

Spinal DAVF presents with gradually worsening sensory disturbances, diffuse back and muscle pain, weakness and sphincter disturbances. Acute onset of symptoms is mostly attributed to spinal haemorrhage either into the subarachnoid space or intramedullary and rarely to venous thrombosis [7, 8]. Spinal DAVF commonly presents with signs and symptoms of progressive myelopathy. Spinal AVM may also present with pain, acute myelopathy, or radiculopathy [9]. Spinal SAH is reported in approximately half of symptomatic spinal cord AVM [10,11] and is frequently accompanied by intracranial signs and symptoms [12].

Case Report

A 3-year-old presented with sudden onset of headaches, weakness of the right leg, photophobia and neck stiffness. On examination MRC grade 2/5 power was noted in the right leg.

Non contrast CT scan revealed blood in the subarachnoid spaces and occipital horns of the lateral ventricles. Cerebral CT angiogram demonstrated no intracerebral aneurysms or AVM. However, blood was noted at the foramen magnum which raised suspicion of a spinal origin of the SAH. (Figure 1).

CT angiogram of the spine revealed a left sided Type IV dural AVF with a feeding vessel entering the spinal cord at the left C6C7 foramen (Figure 2).

A C6-C7 laminectomy was performed followed by a longitudinal durotomy to expose the dural fistula. A temporary clip was applied to the suspected feeding artery and subsequent collapse of the malformation was noted. The artery was then ligated using ligaclips and the temporary clip was removed. Watertight closure of the dura was obtained, and the C6C7 lamina replaced using a microplating system.

Postoperative resolution of leg weakness was noted. However, the patient developed non communicating hydrocephalus as a result of the intracerebral blood, necessitating the need for a VP shunt.

Discussion

Spinal vascular lesions are partially understood and is often described by overlapping terms in the different classification schemes. Incidence is about 4% of primary intraspinal masses, with 80% occurring between ages 20-60 years. Many classification schemes have appeared in the literature since Bergstrand’s first attempt in 1964.

Spinal DAVF have been reported as a rare cause of intracranial SAH. However, mainly patients with cervical DAVF have been described as being indistinguishable from patients with an intracranial source of bleeding [1,
The pathological mechanism of intracranial SAH from a Spinal DAVF remains unclarified. The most straightforward mechanism is migration or extension of subarachnoid blood from the spinal to the intracranial level [14, 15]. Haemorrhage may be caused by venous hypertension when arterialized blood flows via the medullary vein to the valveless coronal venous plexus and radial vein [16, 17]. Another hypothesis suggests that the vein around the midbrain is compressed or stretched by the tentorial incisura when, eg., physical exercise elevates the ICP, which then leads to aggravation of venous hypertension with subsequent rupture of the vein [18]. In patients with intracranial drainage of their Spinal DAVF, the relatively fast venous flow may cause formation of a varix on the draining vessel, which may result in intracranial SAH after rupture [19]. Ascending venous drainage was associated with an increased risk of SAH in patients with CCJ perimedullary and dural AVF [20]. In this light, it is not surprising that in patients with Spinal DAVF, cranial symptoms and signs other than those suggestive of intracranial SAH caused by rupture of a saccular aneurysm have been reported, such as intermittent double vision, slurred speech, and nystagmus [21].

Conclusion

Spontaneous subarachnoid haemorrhage in the paediatric population is an uncommon entity with several possible causes, with a Spinal DAVF being even more uncommon. Clinicians must have a high index of suspicion of an underlying lesion and in today’s practice, the use of available investigations is of utmost importance.

References

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