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Pediatric Arteriovenous Malformations of the Neck: A Case Presentation and Review of Literature

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Abstract **Case Report**

Congenital high-flow AVMs are pathologic connections between arteries and veins anywhere upstream of capillary level. These can either be direct fistulous connections or more commonly contain an intervening nidus—a convoluted network of blood vessels with poorly differentiated endothelial cells. Natural history and symptomatology largely depend on extent and location of the lesion. Venous hypertension as a result of chronic arterialization of the draining veins is a major source of early morbidity. Distal ischemia is a later manifestation. Endovascular treatment has evolved into a mainstay of treatment. Although direct arteriovenous fistulas can be cured by use of proximal occluding devices, AVMs with a nidus require infiltration with a liquid embolic agent delivered super selectively via a coaxial microcatheter-based system. Without this, all attempts at more proximal inflow control are futile. Given their convoluted and evolving angioarchitecture, AVMs readily render themselves to creativity and technical innovation during treatment. Experience and expertise with various therapeutic modalities remain the most important determinants of clinical outcomes.

Keywords: Pediatric Arteriovenous Malformations, Neck, blood vessels, Venous hypertension, chronic arterialization.

INTRODUCTION

Vascular anomalies characterized by the abnormal development or growth of blood and/or lymphatic vessels comprise a spectrum of diseases that have a wide range of complications and varying severity.

In 1996, the International Society for the Study of Vascular Anomalies (ISSVA) adopted and revised the classification system created by Mulliken & Glowacki in 1982 [2]. This has since been considered the official classification system of vascular anomalies. They proposed the division of these anomalies into two major groups: tumors (infantile hemangioma, congenital hemangioma, tufted angioma, Kaposi hemangioendothelioma) and malformations. The vascular malformations can be subdivided into low-flow (capillary, venous, or lymphatic) and high-flow malformations (arterial malformation and arteriovenous fistula) [1].

Arteriovenous malformations (AVMs) are characterized by an abnormal leash of vessels allowing for arteriovenous shunting. They can occur anywhere in the body but are most common in the brain or spinal cord but they can occur anywhere including within the neck [3]. There is direct arteriovenous communication with no intervening capillary bed. They can be congenital or acquired.

Unlike most vascular malformations, AVMs may remain quiescent until puberty and in rare cases into adulthood and are usually triggered by trauma, infection, or hormonal factors [4, 5]. Although many AVMs are asymptomatic, they may alternatively trigger severe pain and/or bleeding. The most common symptoms are pulsation (51.2%), bleeding (41.5%), and pain (29.3%) [4].

In principle, AVMs can effectively be managed by total resection, usually achieved with surgery combined with embolization or surgery alone. In this case report, we present another occurrence of AVM of the neck in a 2-month-old infant along with a discussion on the diagnosis and treatment options.

CASE REPORT

A 2-month-old infant with a congenital cervical mass is observed (Figure 1). The mass is soft to the touch

and pulsatile and it has increased in size since birth. Initially, a cervical ultrasound was performed revealing a hyper-echoic heterogeneous mass that was hypervascularized on color Doppler. A CT scan demonstrated a complex hypodense lesion in the posterior right cervical region measuring approximately 56 x 40 x 24mm. The angiographic images showed a complex network of tangled vessels making up the mass. There was supply predominantly via the vertebral artery which was enlarged as well as a branch from a right external carotid artery. Drainage appeared to be into the right internal jugular vein (Figure 2 & 3). Based on these findings, the history, and the clinical presentations, a diagnosis of AVM of the neck was made.

Figure 1: Cervical AVM

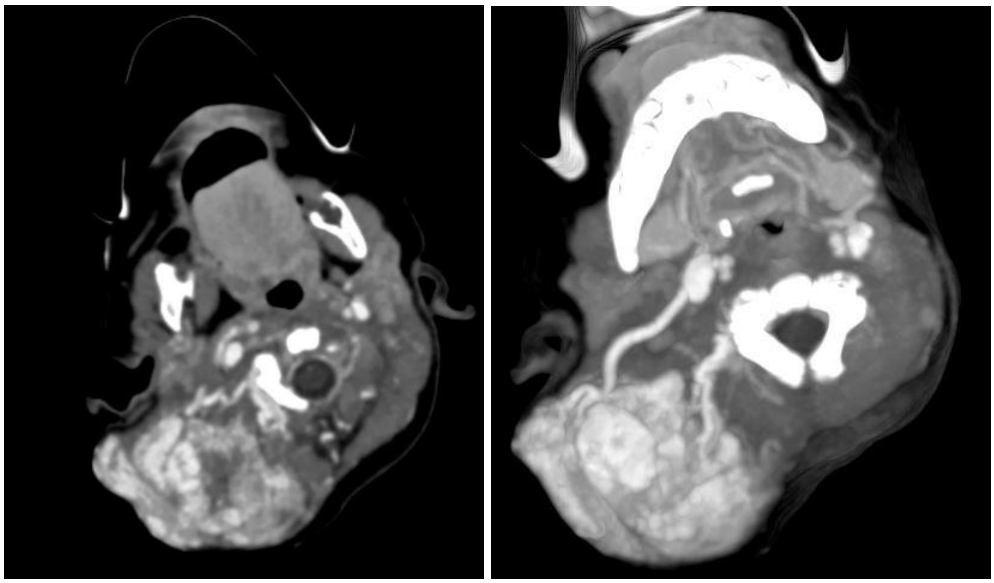

Figure 2: The axial view of the CT angiography reveals a nidus predominantly supplied by the enlarged vertebral artery and a branch from the right external carotid artery. The venous drainage is observed into the right internal jugular vein

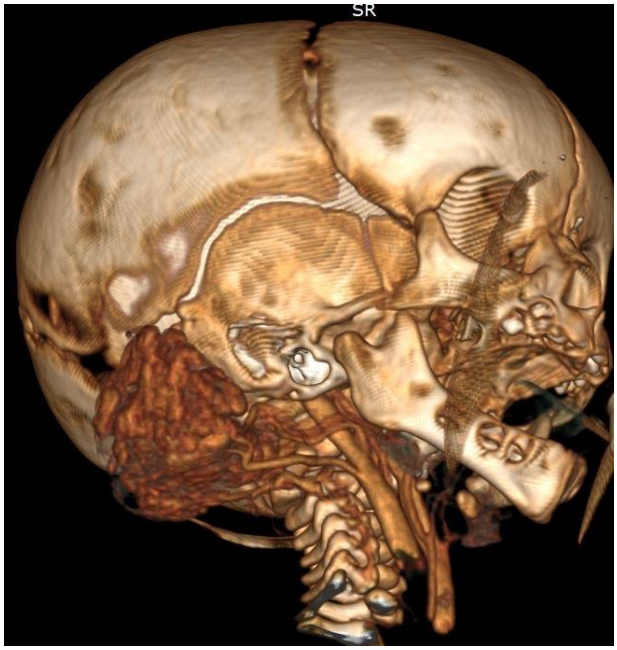

Figure 3: The Standard Volume Rendering Technique (VRT) visualizes the cervical arteriovenous malformation (AVM)

DISCUSSION

Arteriovenous malformations (AVMs) are usually present at birth and associated with clinically significant arteriovenous shunting. The head and neck region, including the brain, is the most common site of involvement. Superficial lesions may raise skin temperature and produce palpable pulsation. In addition, hemorrhage and local tissue ischemia secondary to arterial steal are often seen. The lesions tend to show progression over time due to collateral arterial flow. Most AVMs are sporadic; however, a subset of these lesions are part of inherited syndromes (hereditary hemorrhagic telangiectasia, Parkes Weber syndrome, and capillary malformation/CM-AVM). AVMs have been associated with somatic mutations involving the mitogen-activated protein kinase kinase 1 gene (MAP2K1).

Histologically, the actual arteriovenous shunts are difficult to identify (extensive sectioning is often required). The lesions often demonstrate arterioles, capillaries, and venules within a densely fibrous/fibromyxoid background intermixed with large and tortuous arteries and thick-walled veins. Luminal thrombi and intravascular papillary endothelial hyperplasia are absent, in keeping with the abnormal high venous flow and pressure.

Imaging

Duplex ultrasonography can be an adequate initial screening modality for more focal superficial AVMs. B-mode imaging and color-flow Doppler characteristically reveal a hyper vascular network of dilated tortuous channels, including multiple arterial feeders and venous drainers. The latter may at times undergo aneurysmal degeneration as a result of long-standing arterialization of the venous system. Spectral waveform analysis reveals a high-flow, low-resistance vascular bed. Definitive diagnosis, however, is often made via contrast-enhanced magnetic resonance imaging. AVMs characteristically feature moderate T2 enhancement with interspersed flow voids, which represent foci of high-flow shunting within the lesion.

CT is ideal for bony AVMs, whereas selective angiography is most useful for further investigation of AVMs to identify the specific arterial supply. In the present case, which involved superficial tissues of the head and neck, physical examination and the clinical history were sufficient to determine the clinical diagnosis of AVM. However, for further investigation needed for effective surgical intervention, a CT angiogram was indicated. This was successful in identifying the complex network of tangled vessels making up the mass and the feeding and draining vessels. To further aid in diagnosis, cinematic rendering was employed to create 3D photorealistic representations of the AVM, which helped in better understanding the medical condition's tortuous vascular structure.

Treatment

Treatment aims to obliterate the nidus to prevent lesion growth by stopping the recruitment of new vessels. Small asymptomatic AVMs do not require treatment. Symptomatic AVMs are treated with embolization and sclerotherapy to facilitate safer surgical resection. Ligation of feeding vessels should be avoided as it can lead to collateral circulation and further enlargement. Super selective embolization is rarely successful alone but may be used for palliative care in inoperable AVMs. Surgical resection aims for total excision to prevent recurrence, which is common with

subtotal resection. Overall AVM management is complex and requires a multidisciplinary approach and appropriate imaging for optimal treatment outcomes.

Declaration of Interest: All authors declare no conflicts of interest.

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