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Case Report

A Rare Case of Congenital Malformations of Left Subclavian Arteriovenous Underwent Surgical Resection

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Background: Arteriovenous malformations (AVM) that occur extra-cranial are less common compared to intracranial lesions. The potential for this expansion of AVM throughout life with soft tissue invasion, bleeding, deformity, and functional deficits has caused much attention in terms of management.
Case: A 9-months-old boy is referred to bleeding due to an infected right shoulder ulcer since the age of 1 month. An angiogram performed previously shows right shoulder AVM. It is clear that excision is an appropriate treatment because the location is easily accessible and there is no extension to the thickened area or neck. Histopathology of the lesion showed a lobulated mass consisting of proliferating veinsand arteries of varying sizes, mostly capillary size. The blood vessels were dilated and lined with endothelial cells without any sign of atypia. Part of the lumen of blood vessels contains erythrocytes. The patient has a good prognosis after extensive excision**.**
Conclusion: Congenital subclavian AVM are extremely rare among those extracranial AVMs, and their treatment can be challenging. Surgical treatment may be needed for patient

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INTRODUCTION

Peripheral vascular malformations are rare. Vascular malformations occur most frequently at intracranial sites. This malformation appears from birth and gets bigger as the child grows, especially the involvement of Notch4's expression and signaling.[1] These masses fail to regress and tend to develop rapidly at puberty, pregnancy, or after trauma or intervention. Malformed vessels include lymphatic channels, arteries, veins, capillaries, arteriovena, and a mixture of them with or without a fistula.[2] About 70% of these malformations are complex malformations.[3] AVM can appear anywhere, including extracranials most commonly in the head and neck followed by neck, trunk, and visceral locations.[3,4]

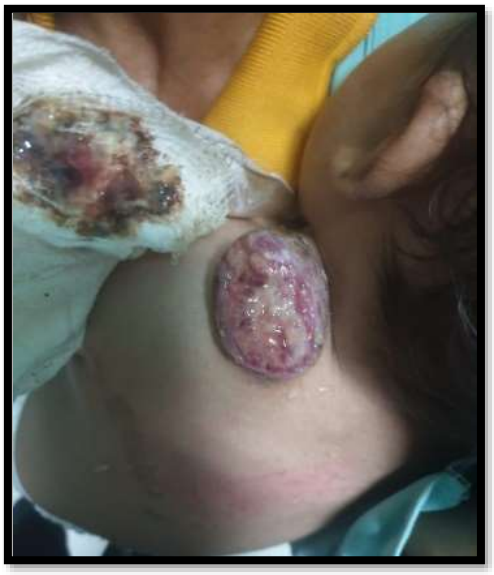
Peripheral arteriovenal malformations cause clinical conditions that vary depending on the surrounding organs and can be life threatening. In areas that are easy to examine such as the limbs, the AVM appears as a soft tissue mass with or without pulsation or thrill.[5] This occurs due to a direct connection between the arteries and veins that bypass the capillary pads. Decreased levels of oxides delivered to tissues are ischemic and tends to ulcerate. Other symptoms can include lymphatic edema, mendantism of the extremities, and excess cardiac return.[6] Diagnosis by angiography can help to determine the appropriate treatment for the patient. Duplex ultrasound can be the initial screening tool in superficial and focal AVMs.[7] The ultrasound image shows a dilated, winding, blood vessel with several main arteries leading to a single venous drain. Other investigations with Magnentic Resonance Angiography (MRA) or Computerized tomography angiography (CTA).[8] CTA imaging produces a better picture of MRA, but this examination is not recommended in infants and children because of the large radiation risk.

The treatment of AVM, depending on the type is extratruncular or truncular. In extratruncular AVM it can be either occlusion or removal of the AVM tissue. One of these actions is expected to be a permanent treatment for the patient. Extratruncular AVMs have to be aggressive because of the easily accessible location. In this case report we will discuss the case of an AVM on the shoulder of a baby boy.

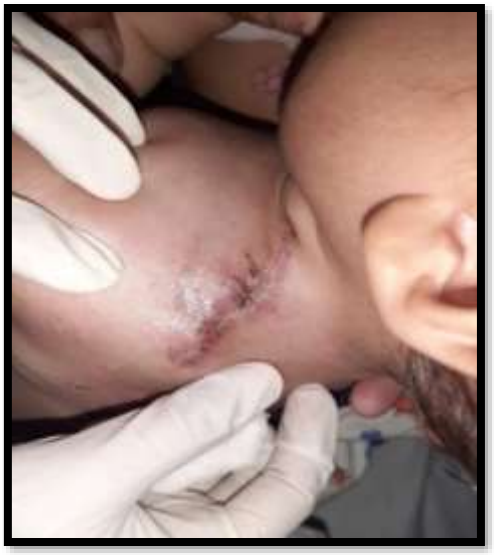
This case was raised because the case of AVM in the shoulder is a case that is rarely reported.

Case Report

A 9 months old male baby presented with a wounded mass on his left shoulder which has been occurring for 8 months. Soft easily bleed mass was palpated without pulsation nor thrill on auscultation. His parents stated that the mass was present from birth and had gradually enlarged during several months. Patient often came to the hospital to treat his recurrent infected wound. His right arm seemed bigger, so the movement of his left arm became less active, Figure 1.



A.

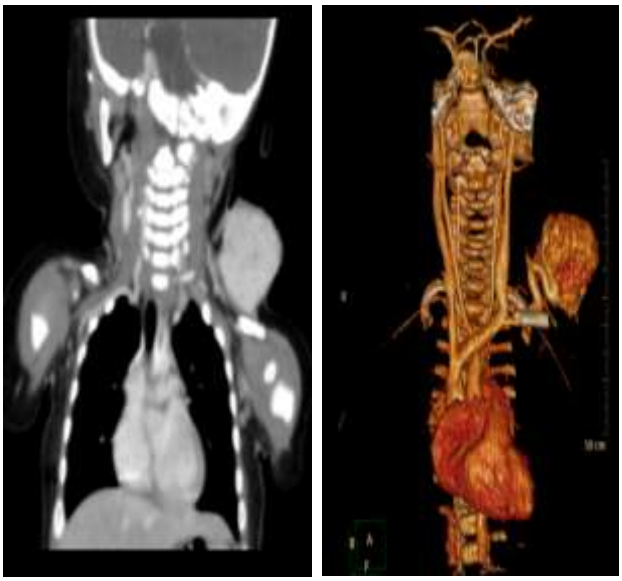


B.

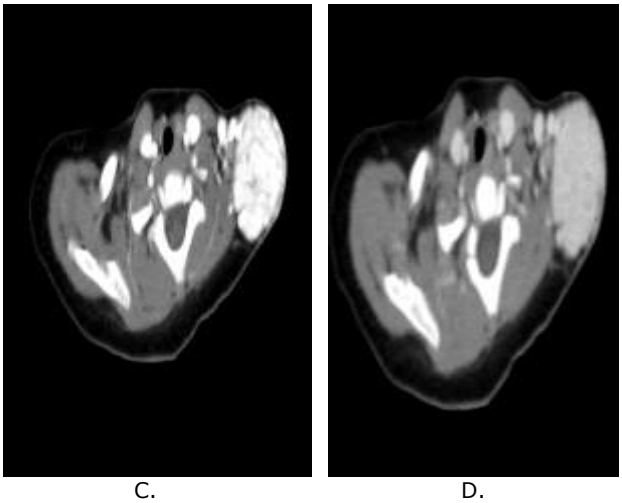
Figure 1. (A). AVM of left shoulder with chronic ulceration. (B) left shoulder after AVM resection.

A Compute topography angiography demonstrated a hyperdense lesion in the left supraclavicular regionwith a size of 4.3 x 3.5 x 3.7 cm. This figure was consistent with an AVM in the supraclavicularregion which receives a main artery from the left subclavian artery junction and drainage to the left external jugular vein, Figure 2.

A wide excision surgery and removal of the AVM were performed and accompanied by reconstruction. The malformation was accessed via supraclavicular incision extending into the posterior shoulder. Dissection was difficult due to a large number of collaterals. The subclavian vein and artery were dissected underneath the clavicle. Feeders from the left subclavian artery junction and external jugular vein were ligated. Proximal control of the subclavian artery was gained at this stage. The sac of the malformation was dissected on the posterior shoulder wall. Upon opening the sac, there was blood loss around 250 ml. The vascular communications to the sac were the closed directly. Following this, the sac was completely closed after disconnecting all the systemic connections.



A. B.



C. D.

Figure 2. (A) CT with coronal view; (B) Reconstruction CT-Angiography showing AVMwith feeding of the left subclavian artery; (C) CTA at the arterial phase; (D) CTA in the venous phase.

Histopathology of the lesion showed a lobulated mass consisting of proliferating veinsand arteries of varying sizes, mostly capillary size. The

blood vessels were dilated and lined with endothelial cells without any sign of atypia. Part of the lumen of blood vessels contains erythrocytes, Figure 3. After surgical procedure the wound dries well.

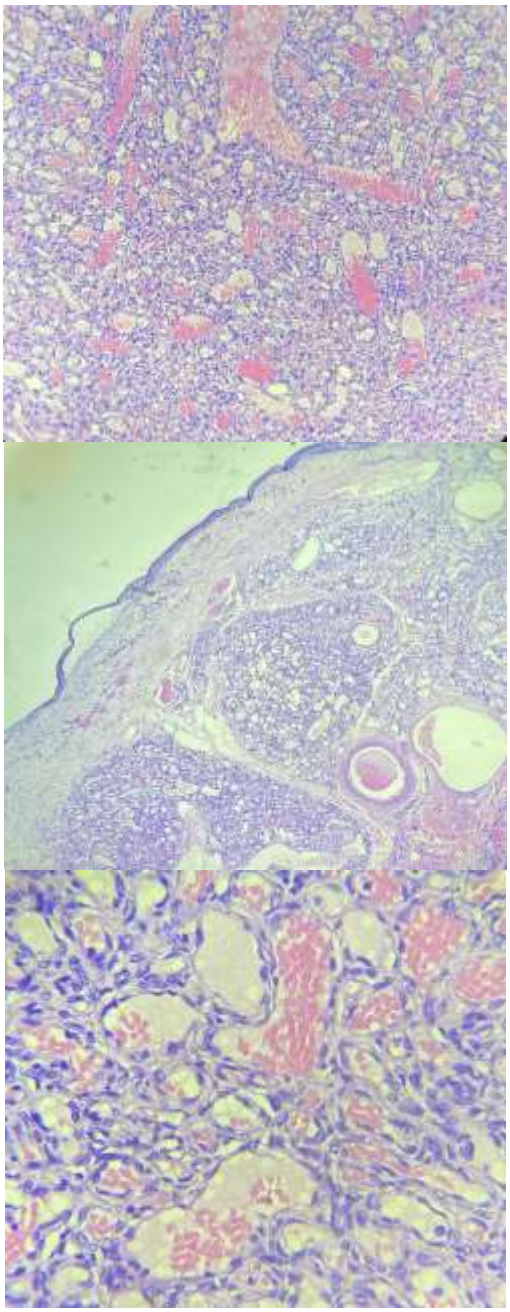


Figure 3. Histopahology of the lesion showed AVM

Discussion

An arteriovenous malformations (AVM) are rare congenital vascular malformations characterized by abnormal arteriovenous connections and high-flow arteriovenous shunting.AVMs are rarely seen in the subclavian artery, and the most common caused are trauma or iatrogenic.[8] Congenital subclavian AVM are extremely rare and their treatment can be challenging.The management of it is a high risk procedure because of the presence of important structures, including the subclavian artery, veins, and nerve plexus.[9]

AVM shows a wide range of clinical presentations. AVMs involving skin and deep soft tissue are characterized by a poorly defined blush or red mass that may be pulsatile, with variably severe arteriovenous shunting, sometimes leading to local tissue necrosis, haemorrhage, or high-output heart failure. Patient with subclavian AVM may suffer from pain, nerve compression, ulceration, hand dysfunction, and spontaneous bleeding. Spotting through proximal arteriovenous fistulas can cause a distal stealing phenomenon that manifests as severe pain, ischemia and discoloration of the fingers.[10]

Management of AVM is challenging, which includes conservative management, surgery and interventional radiology using catheter embolisation or direct percutaneous sclerotherapy.[9,11–14] Treatment of AVM has a high recurrence rate and incomplete treatmentof AVM can make the lesions larger. More invasive treatment may be needed for patients with vascular malformations if they have uncontrollable pain, clinically significant heart pressure, untreated ulcers, functional impairment, or disabilities. Complete resection or occlusion of the nidus is required for full AVM healing. The resection should be as complete as possible because the rate of recurrence is high. In this case, surgical resection was considered to be the best choice for treatment. The nidus of the AVM was completely removed and ligation of feeding artery was done[14]

Surgical resection in the area of the subclavian artery requires a supraclavicularincision with clavicular resection or a thoracotomy.[9,14] The malformation in our case was accessed via supraclavicular incision extending into the posterior shoulder.

Surgical treatment alone can induce massive bleeding during surgery thus most surgeons recommend preoperative sclerotherapy [14] In our case the total blood loss was around 250 ml.

Conclusion

Congenital subclavian AVM are extremely rare among those extracranial AVMs, and their treatment can be challenging. Surgical treatment may be needed for patient with symptomatic manifestation such as recurrent infected wound. Complete resection of the nidus and feeding artery ligation are required for full AVM healing. Massive bleeding is one of complication during surgery.

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Declarations

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Ethical approval: Parent of patient accepted for publish in this rare case

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