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Case Report

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A Case of Neck Avm Supplied By Arteria Lusoria

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Introduction

Classification of Vascular malformation is based on ISSVA classification. Here is a brief outline of vascular malformation classification.

Vascular malformations
• Low flow vascular malformations
• Capillary malformation
• Venous malformation
• Lymphatic malformation
• High flow vascular malformations
• Arterial malformation
• Arteriovenous malformation
• Arteriovenous fistula
• Combined vascular malformation (combination of above)

Symptoms of high-flow lesions correspond to the degree of AV shunting and the involved area of the body. The prevalence of AVMs in the general population range from 5 to 613 per 100,000 people. Sequelae of expanding AVMs with AV shunting may include ischemic changes, indolent ulceration, intractable pain, acute life-threatening hemorrhage, or recurrent intermittent bleeding. Increased cardiac output with subsequent congestive heart failure occurs in less than 2% of cases.

Patients with facial AVMs of the skin and/or facial bones may present with facial asymmetry, gingival hypertrophy, unstable teeth, periodontal bleeding, or skin/mucosal ulcers with secondary infection. Bony AVMs create osteolysis. Distal extremity AVMs may lead to ischemia of the tips of fingers or toes, which is associated with an arterial steal and venous hypertension. Lower limb skin changes resembling brown-violaceous plaques may appear and are known histologically as pseudo-Kaposi sarcomas.

Stage	Clinical symptoms
I	(Quiescence) Skin warmth, discoloration
II	(Expansion) Enlargement, pulsation, bruit
III	(Destruction) Pain, ulceration, bleeding
IV	(Decompensation) Cardiac failure due to volume overload

The left aortic arch with an aberrant right subclavian artery, or arteria lusoria , is the most common aortic arch anomaly, occuring in 0.5 2.5% of individuals. Four vessels arise sequentially from the aortic arch: the right common carotid artery, the left common carotid artery, the left subclavian artery and the aberrant right subclavian artery, which crosses upwards and to the right in the posterior mediastinum. It is usually asymptomatic. When symptomatic, it produces dysphagia lusoria or yspnea andchronic coughing.

Here we have a case report of a vascular malformation that was scanned at Hospital Shah Alam . The case is a arteriovenous malformation supplied by an aberrant right subclavian artery. Mdm S, a 33 years old indian female with 2 years of right base of neck swelling. Schobinger classification 2. No odynophagia. No dysphagia. On examination, there is a soft compressible swelling at right supraclavicular region. Non tender. Upon ultrasound done in Columbia Asia showed vascular lesion at the right base of the neck. Upon CTA neck done on the 28.11.2019, a lobulated right lower neck vascular lesion, superficial to right sternocleidomastoid muscle, suggestive of vascular malformation. On arterial phase there is early venous drainage into the adjacent ectatic right EJV. Unable to clearly identify a feeding artery on current study though a small branch from the aberrant right subclavian artery seen at its inferior aspect. On portovenous phase the ectatic right EJV is homogenously filled up prior to draining into the right subclavian vein. So with regards to thefinding it was an Arteriovenous malformation. The patient had an appointment in December and was counselled for removal of AVM. However patient refused and defaulted subsequent appointments [1,2].

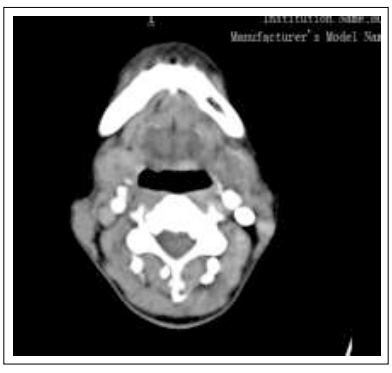

Figure 1

Discussion

Arteriovenous malformations (AVM) develop from an identifiable source vessel called the "nidus," which conducts an abnormal connection of arterial and venous systems. This type of shunt is usually present at birth, but does not become apparent until the first or second decade of life.

In the case above, there is a small branch from the aberrant right subclavian artery supplying the AVM. However patient didn't have dysphagia suggesting no compression from the AVM or the arteria lusoria . No evidence of Komerrells diverticulum or aneurysm. Treatment of an AVM is based on the concept of obliteration of the nidus as this is thought to be responsible for the growth of the lesion through recruitment of new vessels from neighboring regions usually sclerotherapy and embolization. AVMs and AVFs will contain multiple enlarged subcutaneous arteries and veins on grey scale and color Doppler US, with associated low resistance arterial and venous waveforms on pulsed Doppler US. CT angiography is an alternative to MRA that gives comparable images; however, the 3D reconstruction lacks temporal resolution.

Conclusion

The root of the neck is an uncommon location for a soft compressible swelling. Lymph node enlargement can occur in this region but will be firmer in consistency and very well defined. Various swellings, such as intramuscular hemangioma , lymphangioma, lipoma, rhabdomyoma are to be considered in the differential diagnosis. Also, other primary tumours like low grade malignant hemangioendothelioma, hemangiopericytoma and frankly malignant hemangiosarcoma and leiomyosarcoma may be considered in the differential of these vascular lesions. These tumours are all very rare but may enter the differential list if the lesion shows evidence of enlargement or local infiltration clinically or on imaging studies. In addition, AVMs around the subclavian artery are also extremely rare, most are traumatic or iatrogenic; their treatment can be challenging, with a high risk associated with management, because of the presence of important structures, including the subclavian artery, veins, and nerve plexus. Although endovascular treatment is generally the first choice, some patients require surgical resection when endovascular therapy is difficult to perform because of technical or anatomic reasons. In the case of this AVM supplied by arteria lusoria the surgeon has to carefully ligate the feeding vessels first. Also the Schobinger classification is extremely crucial in identifying the sequelae of the AVM with AV shunting. Ideally a follow up of the patient will give us clues of the nature of sequelae as well as local progression of the AVM.

References

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