Rare case of a Congenital Arteriovenous malformation (Park Weber angiodysplasia) around the elbow with median nerve compression

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Citation

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Abstract

A 28 yr old male presented to us with pain and sudden increase in preexisting swelling around the left elbow of one year duration. Examination revealed an arteriovenous malformation with signs of median nerve compression and a fixed flexion deformity of the elbow. Xray revealed erosion of the bone in the upper and the lower end humerus . CT/MR Angiography proved invaluable in delineating the extent of the bony and soft tissue changes. Trauma is one of the factors implicated in the sudden increase of a quiescent AV malformation as in this case.This form of presentation as a high flow congenital arteriovenous malformation around the elbow with osseous changes and nerve compression, we believe is a rare presentation.

INTRODUCTION

Arteriovenous Malformations (AVMs) belong to a group of disorders known as vascular malformations. AVMs are defects of the circulatory system that generally arise during embryonic or fetal development or soon after birth. They consist of masses of abnormal blood vessels.

Peripheral arteriovenous (AV) malformations may present with a plethora of clinical symptoms such as paradoxical emboli, severe hypertension, nerve palsies or pain syndromes. Hemodynamically compromising lesions of the limbs are rare and involve high-flow AV fistulae with marked arteriovenous shunting[1].

Lesions may be located superficially with only minimal arterio-venous shunting or more deeply with significant, high flow, AV shunting.

AVMs consist of a blood vessel “nidus” (nest) through which arteries connect directly to veins, instead of through the elaborate collection of very small vessels called capillaries.

Complete excision of the malformation is required because subtotal resections result in recurrence. Preoperative embolization may be used, followed shortly with operative resection. Simple ligation or embolization is contraindicated for arteriovascular anomalies because rapid collateral arterial flow develops.

CASE REPORT

Presenting complaints -A 28 yr old male c/o pain and increase of swelling around the left elbow. Swelling noticed by the parents since the age of 1 year, slowly progressed up to the age of 15 yrs,remained static until a trivial trauma 6 mths back following which a rapid increase in the size of the swelling was noted. The patient was operated for the same complaints at the age of 15 yrs, operative details not known.

On examination- Diffuse circumferential swelling from the insertion of deltoid to upper third forearm, firm to cystic in consistency

Skin – shiny, multiple engorged veins with thickened walls.

Temp. raised ,erythema of the distal limb present. Operated scar over the medial arm

Limb lengthening of 5 cms noted with significant increase in girth

Palpable and an audible continous machinery murmur. Nicoladinus and Branham sign positive.

Gross restriction of shoulder abduction(60 deg),flexion(40 deg) and external rotation(20 deg).Fixed flexion deformity of the elbow of 30 deg, gross restriction of forearm

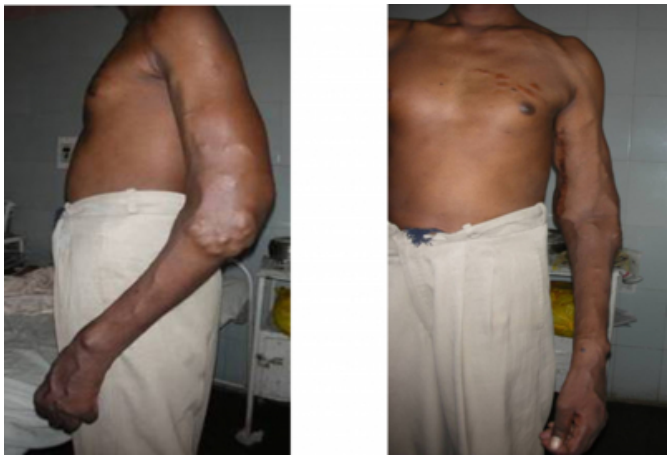
pronation(30 deg).

Paraesthesia in the area of distribution of the median nerve noted, motor system normal..

Schobinger classification- Features of both stage 2 and stage 3(persistent pain).

Figure 1

Photograph 1 Massive swelling around the elbow seen in AP and lateral views



INVESTIGATIONS

X ray -Lytic lesion in head of left humerus

Bony erosions in the lower end of left humerus with pathological fracture.

Ultrasound abdomen and Contrast CT Head – Normal

Colour Doppler -Findings are suggestive of arteriovenous malformation (fistula) at the level of distal third of left arm.

CT/ MR Angiogram confirmed the site and extent of arteriovenous malformation and the presence of a high flow fistula.

Figure 2

Figure 1 X ray elbow

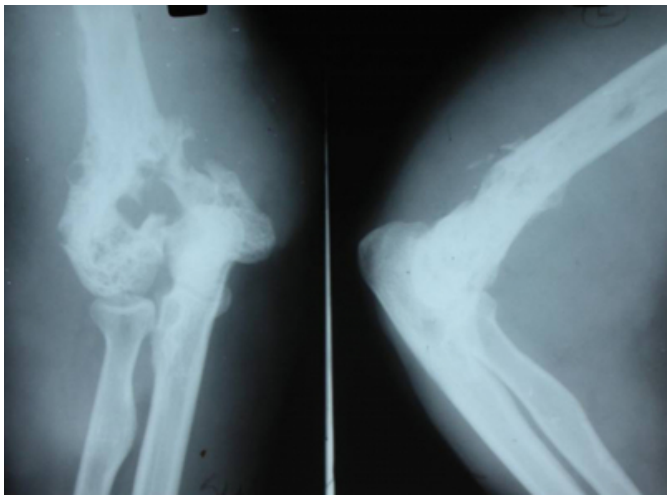


Figure 3

Figure 2 X ray Shoulder joint with proximal humerus

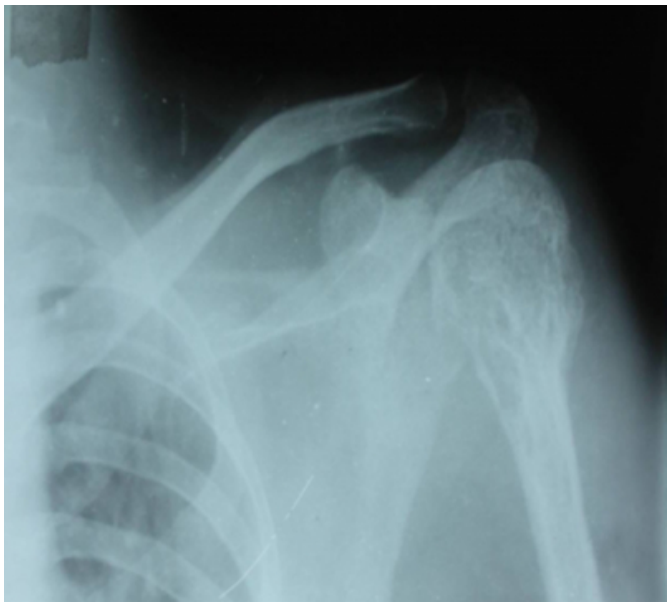
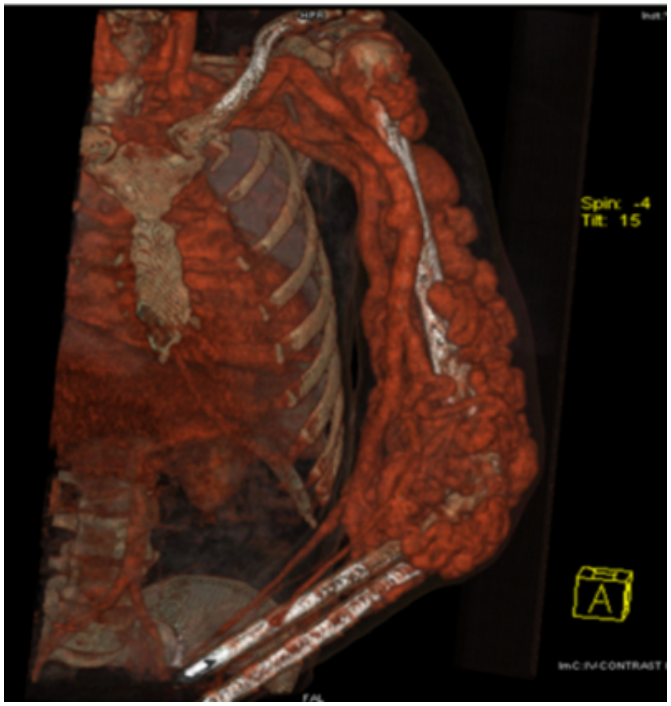


Figure 4

Figure 3 CT Angiogram



DISCUSSION

Arteriovenous malformations are the result of errors of vascular development between the 4th and 6th weeks of gestation and are clearly distinct from hemangiomas, benign tumors with a well-defined life cycle of proliferation and involution. It is believed that expansion is the result of increased blood flow rather than cellular proliferation. Collateral vessel formation is also important in pathogenesis.

Congenital AV fistulas can be divided in to those that primary involve the venous and those that involve the arteriovenous system. Both may or may not have a lymphatic component. AV malformations can be hyperdynamic wherein they cause secondary soft tissue and bony changes or hypodynamic. The one and the soft tissue component might be caused either due to altered haemodynamics or it may represent a primary lesion.

These lesions can remain quiescent for a long time and suddenly increase in size at the time of puberty, infection, pregnancy or trauma[2].

Other organs such as the brain,liver,spleen,lung,kidney,spinal cord can be involved.

Three types of angiodysplasias that involve the musculoskeletal system are the Klippel Trenaunay syndrome, Park Weber and the Servelle-Martorell syndrome.

Cohen et.al studied 47 cases of angiodysplasia and devised a noninvasive method to differentiate between these conditions. This involves taking standard X-rays of the extremities (both sides) which are examined under direct magnification (0.1-01 mm) thus allowing the most exact possible analysis of the skeletal changes. In this way, the Weber syndrome should be suspected if bone prolongation is seen in association with loss of substances from the skeleton. In the Klippel-Trénaunay syndrome, the bone lengthening is not accompanied by bony lesions. In the Servelle-Martorell syndrome bony lesions go hand in hand with limb hypertrophy[3].In Servelle-Martorell syndrome osseous hypotrophy is noted.

AV Malformations are staged by Schobinger classification[4]

Figure 5

Stage	Features
I	Cutaneous blush/warmth
II	Bruit, audible pulsations, expanding lesion
III	Pain, ulceration, bleeding, infection
IV	Cardiac failure

Duplex ultrasound and CT/MR Angiography are the essential noninvasive tests. MRI is a contemporary gold standard for assessing the anatomic status of the AVM. It delineates the lesion as well as the relationship to surrounding tissues and organs, including muscle, tendon, nerve, vessel, and bone. It helps differentiate low-flow and high-flow status.

Transarterial lung perfusion scan (TLPS) was added as a new, less invasive test to measure the extent of arteriovenous shunting of the AVM. The TLPS was implemented not only for the initial diagnosis but also for follow-up assessment of the treated and untreated lesions[5].

Treatment -Combined treatement consisting of highly selective embolization followed by complete resection and reconstruction with local/vascularised pedicle flaps is indicated for symptomatic arteriovenous malformations as in this case.

Conclusion-The recognition of the potential catastrophe that can be caused by an arteriovenous malformation of this magnitude is necessary.A thorough workup towards exclusion of asymptomatic malformations at other sites is mandatory. Treatment as in this case that would necessitate

thorough preoperative evaluation followed by resection and reconstruction of defects is challenging.

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