

INTRAOSSEOUS ARTERIOVENOUS MALFORMATION INVOLVING ELBOW JOINT - A RARE CASE REPORT

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ABSTRACT

Peripheral vascular malformations (PVMs) encompass a wide spectrum of lesions that can present as an incidental finding or produce potentially life- or limb-threatening complications. They can have intra-articular and intraosseous extensions that will result in more diverse symptomology and present greater therapeutic challenges. PVMs are relatively common within the extremities and usually confined to the subcutaneous tissues and muscles. Any misdiagnosis can lead to inappropriate management and treatment. We present to you case of a 40 years old female who presented with complaints of pain in left elbow joint with redness of skin for the last 5 years. Diagnosis of intraosseous arteriovenous malformation was made based on radiological investigations including ultrasound, MRI and angiography.

Key words: Peripheral vascular malformations, intraosseous arteriovenous malformation

Case Report

A 40 years old female who presented with complaints of pain in left elbow joint along with redness of skin for the last five years. There was no previous history of trauma or surgery. Her past medical history was insignificant. Her lab reports were normal except for CRP which was slightly raised. Pain was subsided after taking pain killers. One year back she again developed painful swelling on the overlying skin of left elbow joint along with increased pain. On physical examination there was discoloration of the skin without any bruising (Fig. 1). There was no discharge or sinus tract formation on examination. Her ultrasound Doppler of the soft tissues was done which revealed heterogeneous large vascular channels within the distal end of humerus and elbow joint (Fig. 2).

An initial diagnosis of vascular malformation was made and an X-ray and MRI of elbow which revealed findings of bony erosions by the tortuous abnormal bunch of vessels more marked in the region of distal



Figure 1: Discoloration of skin

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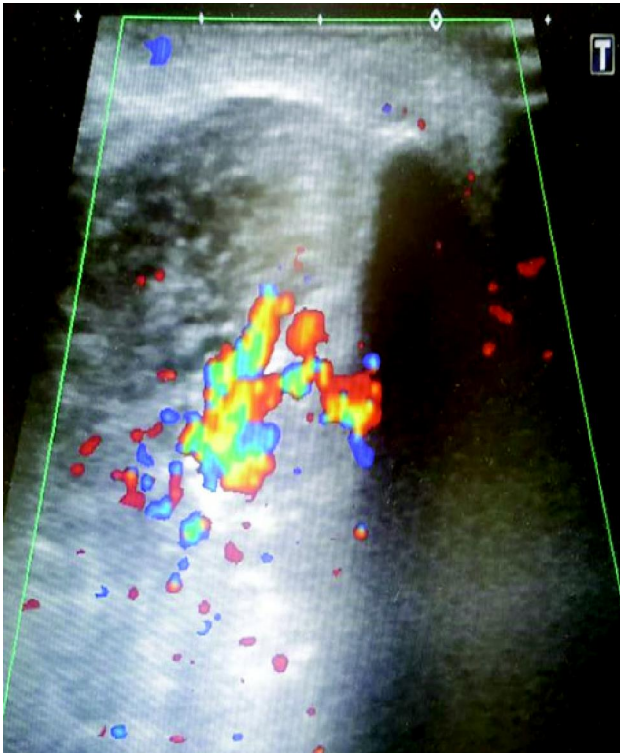


Figure 2: Ultrasound shows bunch of vessels with high velocity flow are seen involving the left elbow joint without surrounding fluid.



Figure 3(a-b): X-ray Elbow AP and Flexed view: There is evidence of multiple lucent areas suggestive of pressure erosions involving the distal end of left humerus and elbow joint.

end of humerus and elbow joint resulting in pressure erosions of the underlying bone. However there was no evidence of any fracture or signs of osteomyelitis (Fig. 3 and 4). Hence a diagnosis of intraosseous



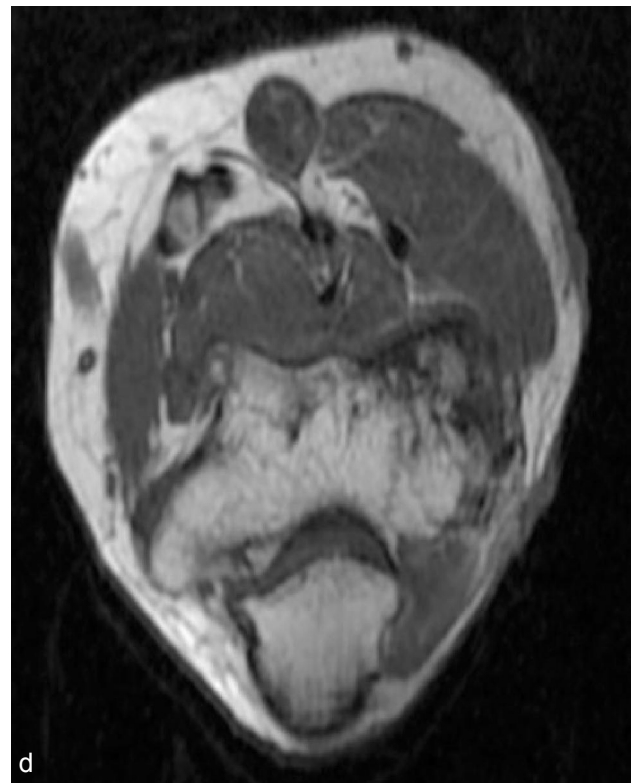


Figure 4(a-d): MRI Coronal, Sagittal and Axial Cuts: There is evidence of multiple tortuous abnormal signal intensity vessels within the substance of bone involving the distal end of humerus and elbow joint. It shows heterogeneous post contrast enhancement. There is no evidence of any fracture. Findings are most likely due to Intraosseous Arteriovenous Malformation.



arteriovenous malformation was made. Therefore an angiogram was performed which showed multiple feeding vessels arising from distal part of brachial, radial, ulnar and common interosseous arteries. These were drained by cephalic and brachial veins. Nidus measured about 2.9 x 2.6 cm (Fig. 5a). Dilated tortuous

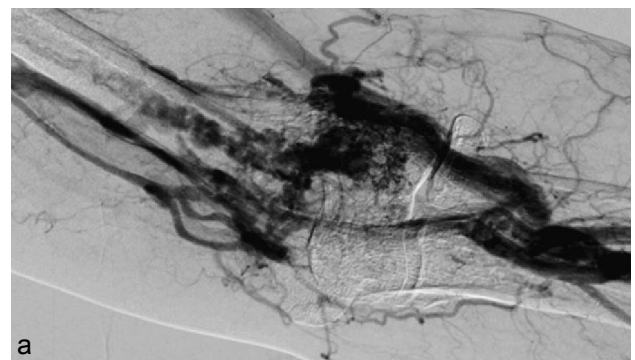


Figure 5(a): Multiple feeding vessels arising from distal part of brachial, radial, ulnar and common interosseous arteries. These were drained by cephalic and brachial veins. Nidus measured about 2.9 x 2.6 cm at the level of distal humerus

arterial channels were also noted in thumb and index finger with early filling of venous channels (Fig. 5b,c). Complete and successful embolization of AVM was performed. No post procedure complications were seen.

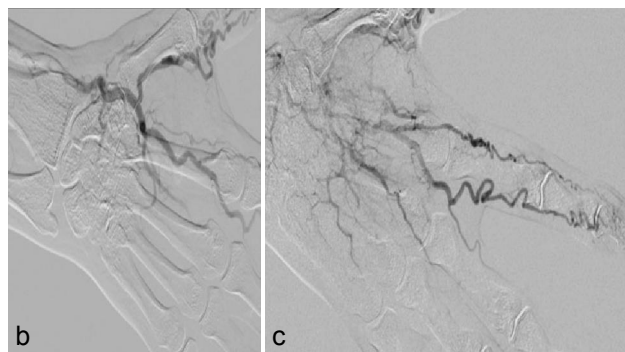


Figure 5(b,c): Few dilated tortuous channels seen at thumb and index finger

Discussion

Mulliken and Glowacki is the most commonly used classification of vascular anomalies.^{1,2} According to this classification; vascular anomalies can either be tumors (principally hemangiomas) or vascular malformations, based on clinical and histological findings. Malformations may also be classified based on flow characteristics.³ High-flow lesions having an arterial component (e.g. AVMs and arteriovenous fistulas) and low-flow lesions refer to capillary, lymphatic and venous malformations.

AVMs are developed between an artery and a vein communicating with each other without any normal capillary network. They are illustrated on imaging by the triad of dilated feeding arteries, enlarged draining veins and early filling of these veins. This was also seen in our case demonstrated on angiography by the presence of a nidus.

Bone involvement by AVMs is extremely rare,⁴ most of which have been reported to occur in the mandible, maxilla and zygoma.⁵ Other skeletal sites, including the tibia, femur, humerus, radius and spine, have also been recounted. Based on these reports, the age group of primary intraosseous AVMs ranges between 7–59 years (mean age of 21); however, all but one of the patients was under 30 years. Intraosseous haemangiomas are being reported in older patients (age group 50s).⁶ The male to female ratio of primary

intraosseous AVM incidence is 4:6, which is similar to that of intraosseous haemangioma incidence. Only 10 cases (including the present case) have been reported the presence of primary intraosseous AVMs in the axial skeleton, and another two cases reported AVM in the vertebra of the spine.⁷⁻⁸

On radiography, most primary intraosseous AVMs of long tubular bones are demonstrated as a central, longitudinal and medullary lytic geographic lesion, occasionally accompanied by sclerotic margins. Three cases showed a hypertrophic nutrient artery groove, appearing as longitudinal radiolucency in the bone cortex. The lesions were found wholly at the diaphysis in cases where the tibia and femur were involved. The other two cases of primary intraosseous AVMs involving the humerus and radius were located in the metadiaphysis. Purely lytic or mixed lytic and sclerotic lesions with a sclerotic margin, distinct from the coarse trabeculation with a corduroy pattern of intraosseous haemangiomas of the spine were seen in the cases of primary intraosseous AVMs of the spine. No aggressive periosteal reactions or associated soft tissue masses were established in the ten reported cases.^{7,8,9}

Our case demonstrates presence of intraosseous AVMs which initially presented like soft tissue hemangiomas. On conventional imaging and MRI, the vascular malformations remained unclear. Angiogram, which showed multiple feeding vessels arising from distal part of brachial, radial, ulnar and common interosseous arteries and were drained by cephalic and brachial veins. Nidus was also seen. Dilated tortuous arterial channels were also noted in thumb and index finger with early filling of venous channels. All these findings confirmed the presence of primary intraosseous AVM.

AVMs are best treated by interventions which include intra arterial embolization, surgery or a combination of both. Ivalon, n-butyl 2-cyanoacrylate (glue), Spongostan, ethanol and microcoils are the most frequently used embolic agents.¹⁰

Conclusion

Peripheral vascular anomalies are rare but catalogue an important series of abnormalities. The symptomatology and imaging appearances of these lesions can

be unfathomable, especially with rarely described intra-articular and osseous AVMs, which can further complicate the diagnosis. Cognizance with the spectrum of imaging findings using different techniques is crucial for the accurate interpretation of AVMs. Radiologists can play a vital role in the diagnosis and treatment of AVMs, whereas digital angiography provides a vascular road map for endovascular treatment. Image-guided percutaneous and interventional treatments are now widely accepted as first-line therapy.

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