“Case Report: Combined extra osseous & intra osseous arteriovenous malformation”

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ABSTRACT:
Arteriovenous malformations are rare congenital lesions caused by faulty development during vascular morphogenesis. There are very few cases reports of combined intra osseous & extra osseous malformations. We report a similar case. We reported 19 year old girl with complaint of swelling in first web space and thumb metacarpal region of left hand since two months which is increasing in size. On clinical examination the swelling in the interosseous space was pulsatile with bruit felt. The veins on radial side of forearm were arterialized. The thumb metacarpal had irregularity on surface & egg shell crackling suggestive of thinning. On x ray the thumb metacarpal showed sieved appearance with thinning of cortex with pathological fracture. On CT angio the whole AV malformation has arisen from a branch of radial artery in anatomical snuff box. At surgery the extrasosseous intrasosseous lesions were found be arising from a branch of radial artery. The whole of extraosseous mass was dissected, the feeder vessel was tied & the mass was removed in toto. Bone was curetted & biconical bone graft from iliac crest was pressfitted in the cavity & fixed with one K wire. Patient was followed regularly. The K wire is removed at three months & mobilization started.

Follow up at one year showed no clinical as well as radiological recurrence. The graft is incorporated well with very good range of carpometacarpal & metacarpophalangeal movements.

Key words: intra osseous, extra osseous, arteriovenous, bone grafting

INTRODUCTION:
Literature search showed the arteriovenous malformations are either intra osseous or extra osseous. After extensive search we could find few cases reports of combined intra osseous & extra osseous malformations. The lesions are congenital but progress with growth spurt. Its timely diagnosis & treatment is necessary to prevent further secondary cardiac effects.

CASE REPORT:
We reported 19 year old female patient presented with pain & swelling in first web space of left hand since three months. The pain was mild, dull aching type, continuous, with no radiation, no diurnal variations. Pain is reduced with analgesics. Pain increased gradually over three months. Patient noticed swelling in left first web space which was having serpentine extensions peripherally on dorsal aspect. The size of swelling also increased over last three months. There was no tingling or numbness distally. No history of similar swellings over body.

On examination there was a diffuse swelling in first web space. Skin over the swelling normal in color, soft in consistency. Swelling was pulsatile & partially compressible. The peripheral veins on dorsum of hand were tortuous, thickened & enlarged. Classical bruit was present on palpation & auscultation.

On palpation of first metacarpal, it was found to enlarged as compared to opposite side. Surface was irregular on
palpable subcutaneous surface, tender with egg shell crackling on deep palpation. Movements at 1<sup>st</sup> carpometacarpal & metacarpophalangeal joints were normal. Plain X ray showed expansive lesion involving first metacarpal shaft with scalloping of edges. The expansion was more towards palmar aspect. The cortex was thinned out. & a pathological fracture is also seen. There was no calcification either in metacarpal or in the soft tissue mass. The epiphysis is intact & the lesion did not cross the physeal plate. The joint on either side appeared to be normal. (Fig) CT scan confirmed the X ray findings. CT angiography was performed (Fig). It showed arteriovenous malformation from radial artery in snuff box area with same soft tissue lesion involving first metacarpal with multiple anastomoses & enlarged veins. Differential diagnosis: While thinking about lesion two things are possible. Soft tissue lesion and bony pathology is different. Looking at radiological picture following differential diagnoses of bony lesion were thought Enchondroma, Aneurysmal bone cyst, Chondrosarcoma, Tuberculosis, chondroblastoma. Plan was done to take biopsy of the bony lesion. At biopsy even under local anesthesia there was lot of bleeding through lesion. A bone piece from wall was removed & sent for biopsy which was inconclusive. Only thing happened was the fracture became complete. Plan was done to explore the lesion. At exploration, the radial artery was exposed in anatomical snuff box. A branch just proximal to princepspollicis artery was found to be feeder vessel for soft tissue lesion & a branch of the same was found to be entering the metacarpal. Metacarpal cortex was having sieve like appearance. There was a membrane like structure which was lining the bone which was hollow enough. On palmar side proximally there was no bone. There was a pathological fracture at the neck due to previous frustrating attempt of obtaining biopsy from bone through a multi-headed blood fountain under local anesthesia. The feeder vessel was ligated & initially the soft tissue mass was dissected by serial ligature of the vessels & the mass was excised. The metacarpal was curetted thoroughly to remove the membrane. A slot was made on palmar aspect by extending the defect. A well shaped bicortical iliac graft was taken & was inserted in the slot under tension. A 2 mm transarticular K wire was put to hold the graft in position. Pressure bandage was given & the tourniquet was released. Stitches were removed at ten days. & K wire was removed at three months. Patient was followed regularly. X ray at one year showed no recurrence with good incorporation of graft with full function of thumb & movements of metacarpophalangeal & carpometacarpal joints.

DISCUSSION

Arteriovenous malformations (AVMs) are rare congenital lesions caused by faulty development during vascular morphogenesis.(1) They consist of abnormal vascular channels called the nidus, comprising of both feeding arteries and draining veins. These are biologically inert lesions that, unlike haemangiomas, contain nonproliferative endothelium. They are present at birth but usually become clinically apparent with growth.(2,3) However some lesions may show aggressive growth that can be triggered by trauma, infection, surgery, puberty, or pregnancy.(4) as in present case puberty was trigger.

They may present at birth as pulsatile soft tissue swelling. In later childhood the swelling may be

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associated with pain on exercise, increased local temperature, sweating & distal ischaemia.(6)
Compressive neuropathy, skin ulceration, uncontrollable bleeding, high output cardiac failure are other known complications.(4,6)

Skeletal changes seen in malformations are seen in up to 34% of cases(3). These changes include bony distortion, bony destruction, hypertrophy, hypoplasia, sclerosis, or osteopenia and primary intraosseous involvement. Intraosseous lytic changes, with a lattice like trabecular pattern (as in present case) are characteristic of high flow lesions with intraosseous involvement. (2). This has been attributed to increased flow in these lesions within the bone causing erosive, destructive, density changes in bone.(3)

Doppler sonography is useful. It shows multiple dilated tortuous channels within the soft tissue. Many of these channels show arterial flow; the draining venous channels show high mean velocity, which indicates a direct communication between arterial & venous systems. (7,8) MRI accurately estimates extent & flow characteristics.(9) with presence of multiple flow voids that indicate high flow on spin echo images. (10) Early enhancement is noted on dynamic contrast enhanced MRI(11).DSA shows direct arteriovenous communications (microfisulas) through the vascular nidus, hypertrophied and tortuous afferent arteries, & early draining efferent veins (12). In present case CT angiography was done to determine the vascular nature of lesion.

The most effective treatment of this lesion is transarterial embolisation.(8) Liquid agents as n butylcyanacrylate (nBCA), ethanol, onyx are ideal (13-15) Percutaneous therapy is an accepted therapy (16). Sclerosants like alcohol, ethanolamine oleate, Polidocanol are used, of which alcohol is most effective. Though literature wise surgery is performed in rare cases that do not respond to embolotherapy. In the presented case as facilities of embolotherapy are not available in our institute and the problem of integrity of bone of metacarpal through bone graft was to be assessed surgery was chosen as option of treatment. Two year followup showed good incorporation of graft with no recurrence.

CONCLUSION:
Combined intraosseous & extraosseous arteriovenous malformations are not very common. Though literature suggests sonography and MRI play important role in complete assessment of lesion & arterial embolisation is treatment of choice; from experience of this case CT angio with surgery also can be chosen as a preferential modality with good end result.

CLINICAL MESSAGE: Treatment of vascular combined soft tissue & bony lesions is challenging. Detailed angiographic evaluation is essential. If facilities of embolotherapy are not available prior to surgery of pathological fracture, primary surgery can give good end result in one setting.
Figure 1: Preoperative X ray.

Figure 2: CT angiography

Figure 3: Postoperative X ray

Figure 4: Follow up Xray at two year

FIGURE 5 : Clinical pictures showing function at two year follow up

REFERENCES:

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