A Rare Case of Distal Humerus Intraosseous Arteriovenous Malformation

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Learning Point of the Article:

Intraosseous arteriovenous is a rare condition which can mimic osteomyelitis/tumor. This is a case report demonstrates that without hesitation it can be treated by extended curettage with good functional results.

Abstract

Introduction: Primary intraosseous arteriovenous malformations (AVMs) are rare and have only been occasionally reported. We herein report a histologically proven case of primary intraosseous AVM in the distal humerus which mimicked an osteomyelitis on radiography.

Case Report: A 27-year-old male presented with complaints of the right elbow pain and stiffness for the past 3 years. He had initially taken treatment at an outside hospital where he was suspected to have right distal humerus osteomyelitis and underwent curettage and biopsy in June 2017. He presented to us in August 2018 with persistent pain even following the first surgery. Repeat radiographs and computed tomography of the right elbow showed features of osteolytic lesion involving the right lateral humeral condyle just adjacent to olecranon fossa (Fig. 1 and 2). Through posterior triceps, splitting approach para-olecranon lesion was resected by intralesional method (burring), and vancomycin-impregnated calcium sulfate (Stimulan) beads were packed in the defect as infection was suspected (Fig. 3). Clinical improvement and restoration of full range of elbow motion were observed on follow-up. Biopsy report surprisingly suggested arteriovenous malformation.

Conclusion: Osteolytic lesion was localized in the lateral margin of the olecranon fossa (Fig. 2a and b). To reach the lesion during surgery was a major challenge as localization of the lesion was missed out by the previous surgery. The occurrence of such a condition is rare and it may take even years to correctly diagnose the disease.

Keywords: Arteriovenous malformation, distal humerus, intralesional extended curettage.

Introduction

Primary intraosseous arteriovenous malformations (AVMs) are rare, accounting for <1% of all primary intraosseous lesions. They are quite variable in their gross and microscopic presentation, yet all can be traced to anomalous development of the primitive vascular system. They may be totally asymptomatic, cosmetically disfiguring, painful, or on rare occasions, cause high-output cardiac failure. Surgical treatment is often unrewarding with recurrence not uncommon. Intra-arterial embolization has shown promising results. Peripheral symptomatic AVMs are uncommon lesion. Arrest or misdirection of the normal developmental pattern occurring at any stage can give rise to anomalous circulatory structures which can be closely correlated clinically and pathologically to the time of the insult. More than one-half of the reported cases of congenital AVMs involve the extremities. Approximately one-third of these occur in the upper extremity versus two-thirds in the lower extremity. While both soft tissue and intraosseous AVMs have identical origins, their clinical presentation and radiographic findings are quite different [1]. The diagnosis and locating the responsible lesion can be challenging and the lesion is often missed or misdiagnosed. We present a rare case of distal humerus AVMs mimicking osteomyelitis/tumor.

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Author's Photo Gallery

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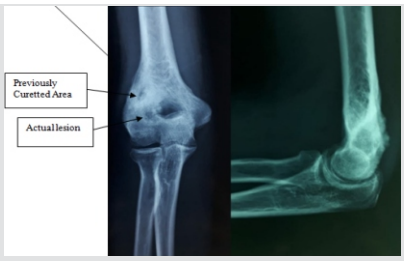

Figure 1: X-ray right elbow anteroposterior and lateral view shown osteolytic lesion involving the lateral condyle of the distal humerus just adjacent to olecranon fossa.

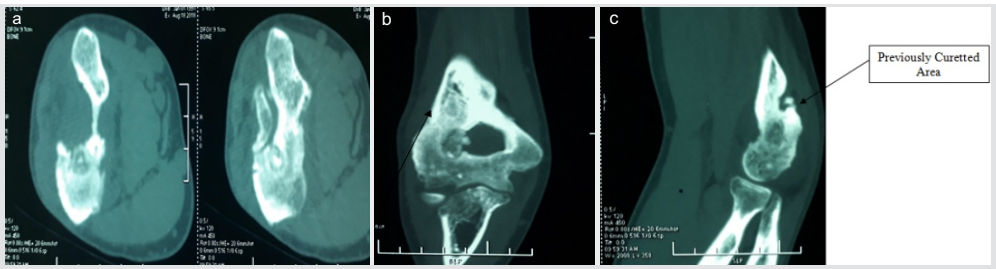

Figure 2: (a and b) An well-defined mixed lytic sclerotic lesion with surrounding sclerosis noted in the distal end of humerus at the lateral margin of the olecranon fossa. (c) The previous biopsy tract which did not lead to the lesion.

Case Report

A 27-year-old male came with complaints of the right elbow pain and stiffness for 3 years. The patient was apparently normal 3 years back. He started developing pain over the left elbow following a trivial trauma which was gradually progressive. The pain was dull aching and aggravated on lifting heavy weights and relieved partially by medications. The patient initially took treatment at outside hospital where he was suspected to have right distal humerus osteomyelitis and underwent curettage and biopsy in June 2017. The curettage was attempted through a lateral approach. As the lesion was in the para-olecranon region, it was probably not reached during the attempted curettage (Fig. 2c), and hence, he had persistent pain even after the surgery.

He presented to us about 1 year later (August 2018) with persistent of symptoms. Examination of the right elbow showed – healed longitudinal surgical scar of length 6 cm present over the lateral aspect of the distal humerus and elbow suggestive of a previous lateral approach, Swelling was present. Tenderness was present over the right elbow and distal humerus. Range of movement of the right elbow was 10–90° (active and passive) further restricted due to pain. Forearm rotations were full. There was no distal neurovascular deficit.

X-ray and computed tomography right elbow showed features of osteolytic lesion involving right lateral humeral condyle just adjacent to olecranon fossa (Fig. 2a and b). On September 05, 2018, through posterior triceps splitting approach lesion, we visualized the olecranon fossa. Preoperatively, olecranon fossa was not breached, and hence, at lateral margin of olecranon fossa was burred to reach the lesion. A bit of red tissue like granulation tissue was identified and curetted out. After extended curettage of lesion with burr, the defect was packed with bone substitute vancomycin-impregnated calcium sulfate (Stimulan) beads as we suspected infection (Fig. 3). Surprisingly, the biopsy was reported as an intraosseous arteriovenous malformation (Fig. 4). Culture of the tissue showed no growth, following surgery, the patient had excellent relief of pain. Postoperatively, the patient was followed up at 1 month, 3 months, 6 months,

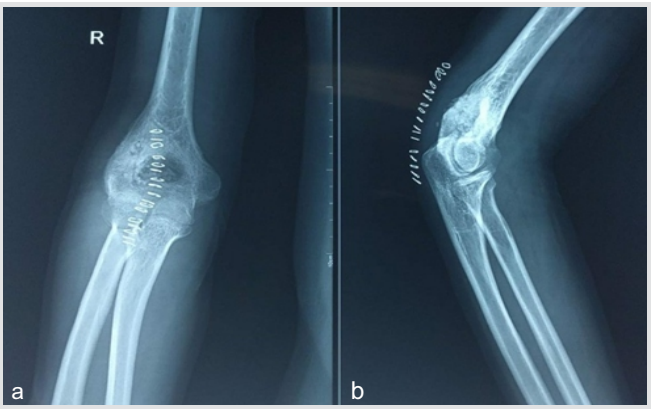

Figure 3: Post-operative X-ray right elbow anteroposterior and lateral views showing lesion excision and antibiotic (vancomycin) impregnated Stimulan beads application at the posterolateral aspect of distal humerus.

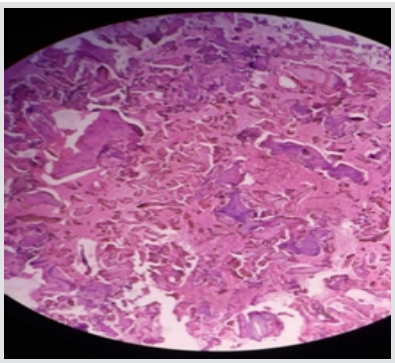

Figure 4: Photomicrographs the intraosseous lesion composed of crowded blood vessels of variable sizes and shapes (Hematoxylin and eosin, ×100); the presence of elastic lamina and fibers in the thickened arterial and venous walls (arrows) (Masson's trichrome stain, ×100).

Table 1: Finding in the repeated cases of intraosseous arteriovenous malformations

Study	Gender/age (Year)	Site	Pain	Pulsatile mass	Radiography	Magnetic resonance imaging	Angiography	Bone scan	Treatment
Present case	F/20	Tibia/diaphysis	No	No	Central medullary lytic lesion with sclerotic margin	Isointense lesion on T1-W, hyperintense lesion on T2-W with a connecting vessel to the normal arterial system of the lower extremity	NA	IU	Surgical resection
Knych *et al*. [2]	M/7	Tibia/diaphysis	Yes	No	Central medullary lytic lesion, hypertrophic nutrient artery groove	Hypointense lesion on T1-W and slightly hyperintense on T2-W	NA	IU	Surgical resection
Matsuyama *et al*. [3]	M/15	Tibia/diaphysis	Yes	No	Central medullary honeycomb lytic lesion, cortical thinning, slight bony expansion	Isointense lesion on T1-W, hyperintense lesion on T2-W with nodular and linear hypointense areas on both T1-W and T2-W	Faint stain of the lesion	NA	Surgical curettage
Katzen and Said [4]	F/28	Tibia/diaphysis	Yes	Yes	Central medullary lytic lesion, hypertrophic nutrient artery groove, cortical defect	NA	Early arteriovenous shunting, hypertrophied nutrient artery	IU	Embolization (Gelfoam)
Savader *et al*. [1]	M/14	Tibia/diaphysis	Yes	Yes	Central medullary lytic lesion	NA	Early arteriovenous shunting	IU	Conservative treatment
	F/28	Humerus/metadiaphysis	Yes	No	Central medullary lytic lesion with thick sclerotic margin	NA	Early arteriovenous shunting	IU	Conservative treatment
	F/22	Radius/metadiaphysis	Yes	Yes	Central medullary lytic lesion	NA	Early arteriovenous shunting	NA	Conservative treatment
Nancarrow *et al*. [5]	F/9	Femur/diaphysis	No	Yes	Small central medullary lytic lesion, hypertrophic nutrient artery groove	NA	Early arteriovenous shunting, hypertrophied nutrient artery	IU	Embolization (coils and Avitene)
Molina *et al*. [6]	M/8	Vertebra L4/lamina	Yes	No	Round lytic lesion with thick sclerotic margin	NA	NA	IU	Surgical resection
Louis *et al*. [7]	F/59	Vertebra T5/posterior element	Yes	No	Expansive mixed lytic and sclerotic lesion with anterior sclerotic margin (on CT), cortical erosion	Isointense lesion on T1-W and hyperintense lesion on T2-W	Feeding artery from T5 intercostal artery	NA	Embolisation (coils) and Surgical resection

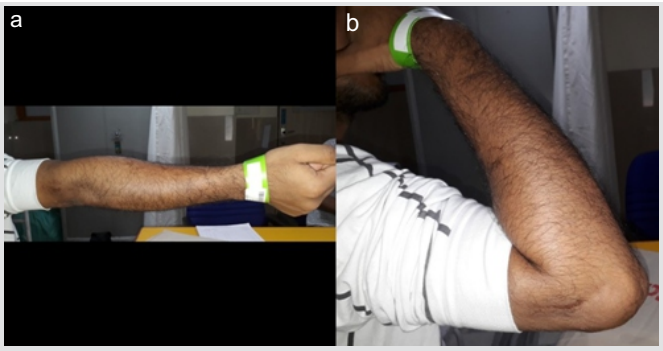

Figure 5: (a and b) Post-operative full range of elbow movements.

and 1 year and he gradually recovered full range of elbow motion with incorporation of bone graft substitutes (Fig. 5).

Discussion

Vascular anomalies are classified as hemangiomas and vascular malformations. The most commonly used system for biological classifications of vascular anomalies is that by Mulliken and Glowacki. This system categorizes vascular anomalies into either tumors (principally hemangiomas) or malformations, based on clinical and histological findings. Vascular malformations may present at birth and enlarge proportionately with the growth of the child. They are the result of errors in morphogenesis and are divided into subtypes such as capillary, venous, lymphatic, arterial, and combined forms, based on the constituent vessels involved. Malformations may be further subcategorized based on flow characteristics. High-flow lesions consist of any malformation with an arterial component (e.g., AVMs and arteriovenous fistulas). Low-flow lesions refer to capillary, lymphatic, and venous malformations. AVMs contain well-formed arterial and venous elements that communicate directly, rather than through a normal capillary network. They are distinguished from other vascular lesions by the triad of dilated feeding arteries, enlarged draining veins, and early filling of these veins. Histologically, the presence of arteries, arterioles, or both as an integral part of the AVMs (demonstrated using Verhoeff's elastic stain) is often used as a diagnostic criterion for differentiating AVMs from hemangiomas. While AVMs often involve the soft tissue and bone, primary intraosseous AVMs that first occur within the bone are extremely rare. Most cases of primary intraosseous AVMs occur in the mandible, maxilla, and zygoma. The occurrence of primary intraosseous AVMs in other skeletal sites, including the tibia, femur, humerus, radius, and spine, has been described in only nine cases to date (Table 1) [1, 2, 3, 4, 5, 6, 7].

Based on these reports, primary intraosseous AVMs occurred at all ages, at the mean age of 21 (range 7–59) years; however, all but one of the patients were younger than 30 years old [1, 2, 3, 4, 5, 6, 7]. Intraosseous hemangiomas are reported in older patients, with the peak diagnosis being made in patients in their fifties. The male-to-female ratio of primary intraosseous AVM incidence is 4:6, which is similar to that of interosseous hemangioma incidence. A total of eight cases (including the present case) reported the occurrence of primary intraosseous AVMs in the long tubular bones of the extremities, and another two cases reported AVM occurrence in the vertebra of the spine [1, 2, 3, 4, 5, 6, 7]. The clinical manifestations of primary intraosseous AVMs are protean. Based on the nine reported cases, painful sensation over the affected area is the most common presentation, occurring in nine of 10 patients, while four in 10 patients present with a pulsatile mass in the area of interest [1, 2, 3, 4, 5, 6, 7]. However, while primary intraosseous AVMs can be associated with skin discoloration, swelling, and bruising, or on rare occasions, cause high-output cardiac failure, it can also be completely asymptomatic.

On radiography, most primary intraosseous AVMs of long tubular bones are described as a central, longitudinally oriented, medullary lytic geographic lesion, occasionally accompanied by sclerotic margins. A hypertrophic nutrient artery groove, appearing as longitudinal radiolucency in the bone cortex, was found in three cases [2, 4, 5]. The anatomic location of the lesions was exclusively at the diaphysis in cases where the tibia and femur were involved [1, 2, 3, 4, 5]. The other two cases of primary intraosseous AVMs involving the humerus and radius arose from the metadiaphysis. Total surgical resection is generally considered the optimal therapy for AVMs due to their tendency to expand and recur with time [8].

During the first surgery, probably, the lesion was not reached as it was done through lateral approach. Hence, the lesion was persistent and the patient was not relieved of his symptoms. We anticipated the difficulty of reaching the lesion through a lateral approach as the lesion was in the lateral margin of olecranon fossa. We initially suspected it to be a subacute osteomyelitis. We were able to reach the lesion and curette it as we had gone through a posterior triceps splitting and not through a lateral approach. Hence, we were able to localize the lesion and remove it intraoperatively. Surprisingly, it turned out to be an arteriovenous malformation.

Conclusion

The diagnosis of arteriovenous malformation is challenging and often misdiagnosed. Extended curettage of the lesion can be done without hesitation to ablate the lesion. Due to the non-specific nature of the presenting symptoms, the condition is often detected late sometimes after years. Proper pre-operative planning, approach, and histology led us to the diagnosis of arteriovenous malformation.

Clinical Message

Osteolytic lesion was localized in the lateral margin of the olecranon fossa. To reach the lesion during surgery was a major challenge as localization of the lesion was missed out by the previous surgery. The occurrence of such a condition is rare and it may take even years to correctly diagnose the disease.

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