

Management of Shaft of Femur Fracture in a Patient with Underlying Arteriovenous Malformation with an Intramedullary Nail

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Abstract

We present a rare case of pathological shaft of femur fracture in an 18-year-old male, with an underlying arteriovenous malformation (AVM). Magnetic resonance imaging showed extensive intraosseous femoral involvement of the vascular malformation, and the canal was deemed to be wide enough for a nail based on the radiograph templates and computed tomography (CT) scan. The patient was given four sessions of stereotactic external beam radiotherapy preoperatively, and closed reduction and internal fixation was performed using a proximal femoral nail without any complications. Radiographic evidence of fracture union was observed at 4 months. Fracture fixation by closed reduction can be either intramedullary fixation or external fixation based on the adequacy of the canal as determined by radiograph and CT scan. Preoperative measures to decrease vascularity such as stereotactic radiotherapy and angiographic embolization can be undertaken to reduce bleeding. A multidisciplinary approach is essential to improve the management of fractures in a patient with AVM.

Keywords: Arteriovenous malformation, femur, fracture, intramedullary nail, radiotherapy

INTRODUCTION

Arteriovenous malformation (AVM) is an abnormal connection between arteries and veins, bypassing the capillary system. AVMs involving bones present a significant therapeutic challenge, if the involved bone sustains a pathological fracture, due to the high risk of bleeding with any kind of invasive procedure, and the poor quality of the underlying bone.^[1] This report presents a case of a pathological shaft of femur fracture with extensive intraosseous AVM, which was successfully treated with closed reduction and internal fixation using a proximal femoral nail.

CASE REPORT

An 18-year-old male presented with pain, swelling, and deformity in his left thigh after a trivial fall. Examination of the left lower limb revealed swelling around the left thigh with

associated abnormal movements. Subsequently, radiography was performed, which confirmed a pathological shaft of femur fracture [Figures 1]. Common differentials of such lesions as per radiographs would include (a) infections, (b) primary bone tumors, (c) endocrine disorders, (d) iatrogenic-like radiation, and (e) secondary bone tumors.

The case in the present study was a known case of congenital vascular malformation involving the left thigh for which he had received four doses of intralesional bleomycin-based

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sclerotherapy. Further evaluation with contrast-enhanced magnetic resonance imaging (MRI) revealed multiple serpiginous and septate lesions with multiple microarterial shunts, phleboliths, and extensive intraosseous involvement. The clinical features of a mass without increased local temperature or thrill with the above-mentioned MRI findings were consistent with low-flow vascular malformation [Figure 2]. It was thus planned to manage it as a low-flow variety of AVM due to the clinical and radiological picture.

Preoperatively, the patient was given four sessions of stereotactic external beam radiotherapy. It was decided to proceed with closed reduction and intramedullary fixation without reaming using a cephalomedullary implant as the canal was deemed to be wide enough for a nail based on the radiograph templates and computed tomography (CT) scan. A backup of a vascular surgeon was ready, and intraoperative armamentarium included a distractor set to

negate any difficulty in closed reduction, external fixation limb reconstruction system, and Ilizarov ring fixator sets as an alternative in case of intraoperative difficulty with intramedullary implants. Two units of packed red blood cells were transfused preoperatively as the patient was found to be anemic. Closed reduction and internal fixation was done on a fracture table under fluoroscopic guidance as planned using a proximal femoral nail [Figure 3]. The procedure was uneventful and did not result in any significant blood loss. The patient was serially followed up to 1 year. The lesion regressed clinically with symptoms of pain reduced, swelling reduced significantly, and the fracture united uneventfully, which was confirmed on radiographs [Figure 4].

DISCUSSION

AVMs can be classified as low-flow or high-flow malformations for management purposes based on clinical, Doppler, and radiological features. In the case of AVMs, MRI has become a mainstay in diagnosis with support from clinical examination and Doppler studies, thus avoiding biopsy, which could carry the risk of bleeding.^[2,3] In our case, we had features of vascular malformation, clinically as well as on MRI. Accurate diagnosis of the lesion is paramount to its effective management. Any discrepancy in the clinicoradiological picture would mandate a biopsy for an accurate diagnosis.

With the growth of the AVM, increased blood flow through the malformation leads to osteopenia and bone destruction, which likely was the cause of the pathological fracture. There have been very few reports of similar cases in the past. Jończyk *et al.* had described a similar case where a pathological subtrochanteric femur fracture with an underlying AVM was managed with an intramedullary nail, in which the canal was deemed adequate for nailing.^[1] Takeuchi *et al.* had managed a similar case with a circular external fixator as their attempt of closed reduction failed.^[4] Others have described four cases of femur fracture in the setting of Klippel–Trenaunay syndrome,



Figure 1: Preoperative anteroposterior (a) and lateral (b) radiographs of left femur fracture with patchy mixed lytic sclerotic bony destruction with periosteal reaction and bony ingrowth into the medullary canal

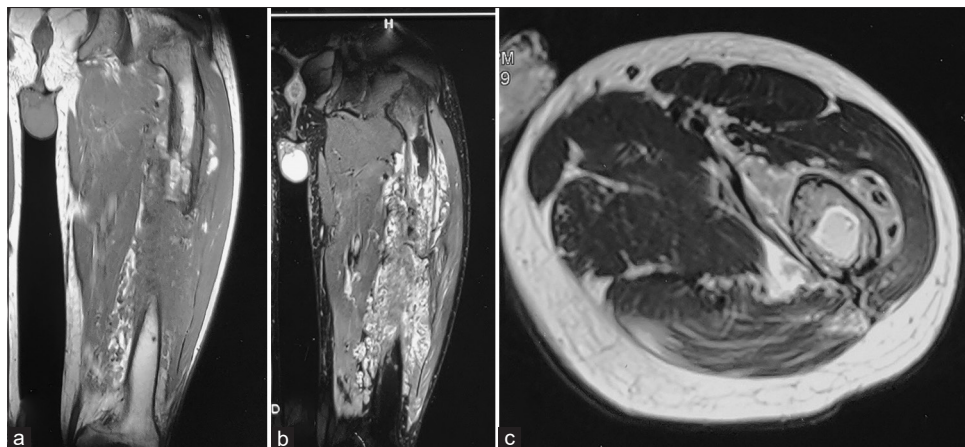


Figure 2: Coronal T1-weighted reduced signal intensity (a), coronal T2-weighted enhanced signal intensity (b), and axial section (c) of magnetic resonance imaging showing intraosseous involvement of vascular malformation, T1-weighted reduced signal intensity and T2-weighted enhanced signal intensity with extensive serpentine septate lesion extending into the bone, with multiple minor feeding vessels to be seen as a signal void suggestive of a low-flow type of vascular malformation



Figure 3: Immediate postoperative anteroposterior (a) and lateral (b) radiographs showing fracture fixation with the proximal femoral nail with satisfactory alignment and reduction

which is also characterized by abnormal arteriovenous shunting, apart from port wine staining of the skin and tissue hypertrophy, and one case with Servelle–Martorell syndrome.^[5-9]

Attempts at nonoperative treatment in view of the high risk of bleeding can result in delayed union or nonunion due to the slow healing rate in such a group. Gibbon *et al.*^[9] conservatively managed a femoral fracture in a patient with Servelle–Martorell syndrome, which resulted in nonunion. Nahas *et al.*^[6] also avoided operative treatment of femur fracture in a case of Klippel–Trenaunay syndrome, resulting in nonunion.

Minimally invasive closed reduction and nailing can be the preferred treatment, especially in a femur fracture, provided the canal is wide. Intramedullary fixation may be difficult if the canal is narrow, in which case closed external fixation may be considered. Open reduction should be avoided in these cases. Takeuchi *et al.* attempted to perform a closed reduction in a femoral fracture with AVM, but failed.^[4] They attempted to perform open reduction, which resulted in severe hemorrhage, following which the wound was closed, Taylor Spatial Frame was applied, and an acute correction was performed.

Even the treatment of such fractures by closed techniques may be complicated by hemorrhage. A 42-year-old female patient previously diagnosed with Klippel–Trenaunay–Weber syndrome, who had sustained a left femoral shaft fracture, was operated with a closed, locked intramedullary nailing procedure by Tsaridis *et al.*, who required multiple blood transfusions.^[8] Our patient had been subjected to four sessions of stereotactic external beam radiotherapy to reduce the vascularity of the lesion.^[10] Other measures such as angiographic embolization of the feeding artery can be done to reduce bleeding.^[11]

The management of a fractured bone with an underlying AVM requires elaborate preoperative planning, expert advice of senior consultants, and interdepartmental consultations.



Figure 4: Postoperative anteroposterior (a) and lateral (b) radiographs at 4 months showing fracture union

Minimally invasive surgery is the preferred modality, especially for a femur fracture. Fracture fixation by closed reduction can be either intramedullary fixation or external fixation based on the adequacy of the canal as determined by radiograph and CT scan. Preoperative measures to decrease vascularity such as stereotactic radiotherapy and angiographic embolization can be undertaken to reduce bleeding. Overall, a multidisciplinary approach improves the outcome of patients with pathological femur fracture with underlying AVM.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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