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Joshi Prateek C

MBBS Assistant Professor
Department of Orthopaedics,
B. J. Govt. Medical College &
Sassoon General Hospitals,
Pune, Maharashtra, India

Chhattani Tushar M

MBBS Assistant Professor
Department of Orthopaedics,
B. J. Govt. Medical College &
Sassoon General Hospitals,
Pune, Maharashtra, India

Shreesha Parthaje

MBBS. Department of
Orthopaedics, B. J. Govt.
Medical College & Sassoon
General Hospitals, Pune,
Maharashtra, India

Rishi Raj

MBBS. Resident Doctor,
Department of Orthopaedics,
B. J. Govt. Medical College &
Sassoon General Hospitals,
Pune, Maharashtra, India

Corresponding Author:**Joshi Prateek C**

MBBS Assistant Professor
Department of Orthopaedics,
B. J. Govt. Medical College &
Sassoon General Hospitals,
Pune, Maharashtra, India

Amputation of an unsalvageable post-traumatic limb in a young Haemophiliac: A multi-faceted clinical dilemma

Joshi Prateek C, Chhattani Tushar M, Shreesha Parthaje and Rishi Raj

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Abstract

A twenty-nine-year-old male presented to the emergency ward of our institute with a history of having been involved in a road traffic accident with a heavy motor vehicle (HMV), 18 hours ago, with trauma to his left lower limb.

Arteriovenous Doppler evaluation of the affected limb confirmed devascularization, and radiographs showed fracture of the femur in supracondylar region and tibial diaphysis with significant soft tissue injury (Letts and Vincent type D floating knee).

Due to the prolonged ischemia time of 18 hours, the Mangled Extremity Severity Score (MESS) of 10, and the contaminated nature of the wound, it was decided to proceed with Guillotine above-knee amputation.

However, a detailed history-taking revealed the patient to be a sufferer of Hemophilia Type A (Coagulation factor VIII deficiency). The patient was stabilized hemodynamically, and supplemented with Factor VIII following which the amputation was done successfully. This case report documents his perioperative and postoperative course in the hospital, ending with his discharge with a small wound healing by secondary intention.

Surgical procedures in hemophiliacs have always presented as clinical dilemmas, with increased blood loss, poor tissue perfusion, and infective events adding to the postoperative complications.

Lower limb amputations in sufferers of hemophilia are very rare, with less than fifty cases being found in the global medical literature. A meta-analysis of said cases reveals the vast majority to have been due to infective indications or for management of hemophilic pseudo-tumours, and post-traumatic above knee amputations are virtually unheard of in such a clinical scenario.

The presence of a traumatic vascular insult in this case, provided a rare clinical predicament, and the successful management of this Catch-22 situation makes for a unique situation.

Keywords: Amputation, hemophilia, fracture, trauma, vascular injury, Factor VIII

Introduction

Haemophilia A, also called classical hemophilia, is a condition resulting from the congenital deficiency of coagulation factor VIII, which results in excessive haemorrhage, either spontaneous or in a post-traumatic setting. With an incidence of 1 in 5000 males and spanning 80% of hemophilia cases, it is the most common of all inherited disorders of coagulation^[1].

It can be graded into three clinical types based on the severity of presentation, as below^[2]:

Table 1: Show the Grade Factor Percentage of Normal

SR.	Grade	Factor Level (Iu/MI)	Factor Percentage of Normal
1.	Mild	<0.01	<1%
2.	Moderate	0.01 – 0.04	1 – 4%
3.	Severe	0.05 – 0.40	5 – 40%

Severe cases tend to present with episodes of spontaneous haemorrhage, whereas mild to moderate variants bleed excessively in a traumatic setting.

Compound fractures are a common occurrence in recent times with an incidence of 30.7 per 100000 population per year. High-energy road traffic accidents form a large part of the demography of these patients with a proportion of 34.1%. Young males are frequently affected^[3].

The constellation of compound polytrauma in a patient presents a unique clinical dilemma in terms of management as well as prognostication.

Case Details

A young male patient aged 29 years, presented to the emergency ward of our institute with a history of a road traffic accident eighteen hours ago. The patient had sustained severe, high-energy trauma to his left thigh and leg and had a 3x5 cm bleeding wound on the anteromedial aspect of the leg in proximity to the shin of the tibia.

Osseous examination revealed fractures of the left distal femur near the supracondylar region, and diaphyseal fracture of the ipsilateral tibia. The patient showed hemodynamic signs of evolving hemorrhagic shock, with tachycardia (heart rate 140 bpm), tachypnoea (respiratory rate 22cpm) and hypotension (80/60mmHg). The patient was drowsy but obedient to verbal commands, with a Glasgow Coma Score (GCS) of 13/15.

A complete hemogram demonstrated a hemoglobin of 9.2 against a hematocrit of 24.2, and depressed counts of erythrocytes. Platelet counts were 106,000, and there was leukocytosis with raised erythrocyte sedimentation rate (ESR).

Further history taking revealed the patient to be a sufferer of mild to moderate Haemophilia A, and prior records of Factor VIII supplementation were unavailable.

Radiographs of the limb corroborated the findings, confirming a Letts and Vincent type D floating knee.

Based on the clinic-radiologic profile and with the hematological investigation reports, the patient was managed as a case of polytrauma with impending Systemic Inflammatory Response Syndrome (SIRS), and stabilized in

the emergency room itself with bilateral broad-gauge intravenous access, Foley’s urinary catheter, oxygen and fluid supplementation, and splintage to the fracture after cleaning and dressing of the wounds.



Fig 1: Lateral radiographs of the distal femur and proximal tibia.

On clinical examination, the affected lower limb was found to be cold and insensate from the mid-thigh distally till the toes, with the popliteal artery, anterior and posterior tibial arteries, and dorsalis pedis artery not clinically palpable.

Spontaneous movements were absent from the limb. The patient was subjected to an immediate arteriovenous Doppler examination which demonstrated absent colour flow in the three distal vessels, and minimal monophasic waveform in the proximal popliteal artery.

Based on the trauma time of 18 hours, the Mangled Extremity Severity Score was calculated to be 10.

Table 1 – Mangled Extremity Severity Score Index.			
Type	Characteristics	Lesions	Sutures
Group of skeletal lesions and soft tissue			
1	Low energy	Sharp wound, simple closed fracture, projectile low-caliber firearm	1
2	Medium energy	Multiple or exposed fractures, dislocation, moderate crush injury	2
3	High energy	Explosion gunshot wound from a high-speed firearm	3
4	Massive crushing	Fall from a tree, train accident, smothering	4
Shock group			
1	Hemodynamically normotensive	Stable blood pressure	0
2	Transient hypotension	Unstable pressure, but responding to intravenous fluid	1
3	Prolonged hypotension	Systolic pressure of < 90 mmHg and responding to intravenous infusion of fluid only in the operating room	2
Ischemic group			
1*	Absence	Pulse without signs of ischemia	0*
2*	Mild	Pulse reduced without signs of ischemia	1*
3*	Moderately	No pulse on Doppler imaging, prolonged capillary refill, paresthesia, decreased motor activity	2*
4*	serious	Pulseless, cold limb, which is paralyzed and numb without capillary refill	3*
Age group			
1		< 30 years	0
2		30–50 years	1
3		> 50 years	2
*Multiply by 2 if ischemia persists for > 6 hours. Limbs with scores of 7–12 points usually require amputation. Limbs with scores of 3–6 points are usually viable.			

Fig 2: Mangled Extremity Severity Score (MESS)

In view of the MESS and the non-salvageable nature of the limb, it was decided to take up the patient for a Guillotine above knee amputation, and the decision of primary closure versus proceeding with an open stump, was deferred to the intra-operative phase.

In consultation with internal medicine specialists, the Factor VIII correction was provided preoperatively using the following formula^[4, 5].

Required dosage in IU = Desired factor level (%) x Weight (kg) x 0.5

The patient was given inj. Factor VIII as a 2000 IU bolus 30 minutes prior to the incision, and followed up with an infusion of 1000 IU B.I.D. over the postoperative period.

With the patient hemodynamically stabilized, he was taken up for Guillotine above knee amputation. Intraoperatively, the supracondylar femur fracture being closed at presentation, no contamination of the soft tissue was found, and the decision was taken to primarily close the amputation site.



Fig 3: Evidently clean wound, postoperative day 1.

Over the course of the next seven days, the dose of Factor VIII infusion was titrated with reference to serial estimations of activated Partial Thromboplastin Time (aPTT) assay.

The patient was stabilized on a reduced bis-in-die dosage of Factor VIII; however, on the seventh post-operative day the patient developed local inflammatory symptoms and spikes of pyrexia and underwent an open debridement of the stump.



Fig 4: The opened wound, with a large clot in the posteromedial quadrant, presumably the nidus of infection.

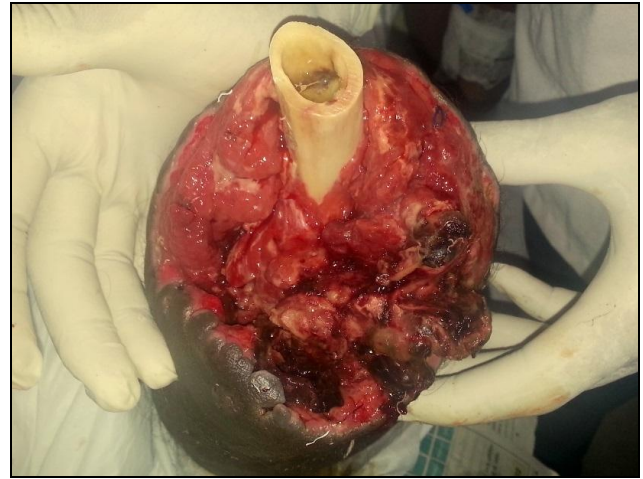


Fig 5: Post debridement status, with the coagulum having been removed en masse. The infected medullary canal was curetted and bone was revised after the photograph.

The patient underwent one more radical debridement and gradual titration of Factor VIII supplementation, along with pus-culture directed choice of antimicrobial agents. With the rapidly contracting wound and healthy granulation tissue, the option of closure was not reconsidered and the wound was allowed to heal by secondary intention.



Fig 6: Clean wound with healthy granulation tissue at 8 weeks postoperative.

Discussion and Review of Literature

Though Hemophilia is one of the very common bleeding disorders in our part of the world, and one of the commonest congenital haemorrhagic diseases known, its coexistence with other conditions can present with unique and challenging clinical scenarios.

The incidence of high-energy road traffic accidents and the frequency of Haemophilia in the general population favours a likelihood of the two coinciding, however, literature related to such scenarios is rare.

Severe trauma to the lower extremity is a common cause of amputation as an end-result, especially when associated with severe soft tissue loss and wound contamination^[6]

Poor outcomes of amputations and limb reconstructions are gauged by the Sickness Impact Profile, a patient-reported outcome measure of multidimensional nature. Poor socio-economic strata, low levels of education, poverty, and major complications all reflect on the SIP to bode a less favourable

outcome, as seen in the patient under discussion [7]. In addition, in a post-traumatic patient with evolving systemic inflammatory response syndrome (SIRS), the values of Factor VIII as obtained on laboratory testing may be subject to variations secondary to elevated and fluctuant levels of epinephrine, fibrinogen, and other acute inflammatory mediators, making clinical correlation a challenge [8].

Among the very first recorded cases of post-traumatic amputations has been cited in 1932, from Nashville, Tennessee, an upper limb amputation for a fractured distal humerus complicated by gas gangrene [9]. Similarly, an above-knee amputation in a hemophiliac was reported for a young male with compound diaphyseal tibia fracture, by Schuster *et al* in 1948 [10].

A tabulated review of literature in this scenario, is as below.

Author	Indication For Amputation	Presence of Infection Preoperative	Open or Closed Amputation	Postoperative Infection	Bleeding Postoperative	Wound Healing Time	Factor VIII	Results
Biggs and MacFarlane	Infected pseudotumor left tibial area (knee disarticulation)	Yes	Closed	Yes	Yes	Time not specified	0 per cent	Survival
Biggs and MacFarlane	Infected hematoma right calf, gangrene foot, BK	Yes	Closed	Not specified	Not specified	3 mos.	0 per cent	Survival
Biggs and MacFarlane	Pseudotumor, left thigh, AK (same patient as above)	Not specified	Closed	Not specified	Recognized tenth postop. day	3 mos.	0 per cent	Survival
Marder and Shulman	Non-union tibial fracture, right-knee disarticulation	Not specified	Closed	Yes	Recognized seventh day	6 mos.	?	Survival
Lewis and associates	Pseudotumor left thigh, AK	No	Closed	Recognized fifth day	Recognized fourteenth day	12 mos.	0 per cent	Survival
Albright and associates	Osteomyelitis right femur, hip disarticulation	Yes	Closed	Not specified	Sixteenth day	6.5 mos.	?	Survival
Pappas and associates	Infected hematoma right thigh, osteomyelitis right femur (pseudotumor), hip disarticulation	Yes	Closed	Not specified	First day and fifth day	3 mos.	23 per cent	Survival
Miles, Von Kaula, Von Kaula	Left AK, osteomyelitis left tibia and fibula	Yes	Closed	Recognized twenty-eighth day	Seventh day small amount twenty-eighth day wound dehiscence and bleeding	3.5 mos.	Not specified	Survival
Schuster	Fracture left fibula, gas gangrene, left AK	Yes (gas gangrene septicemia)	Closed	Second day (stump and septicemia)	Eighth day	Incompletely healed at 8 mos.	Not specified	Survival
Egeberg	Pseudotumor left thigh, hip disarticulation	Not specified	Not specified	Not specified	Sixth day	Not specified	Less than 1 per cent	Survival
Abell and Bailey	Pseudotumor left thigh, AK	Yes	Closed	Immediate	Sixth day	Not healed	Not specified	Death 2 mos. post-amputation
Staas and associates	Infected hemarthrosis, osteo-	Yes	Closed	10 days postop.	10 days postop.	12 mos.	4 per cent	Survival

As evident from the above, majority amputations in haemophiliacs were for the indications of either a hemophilic pseudotumour, or for infection. One case of knee disarticulation following distal tibial infected non-union, has been reported in 1970 by Marder and Shulman *et al*. [11]

Many of the above cases also appear to have been complicated by either postoperative stump sepsis, or septicemia.

Minor surgical procedures such as lipoma excisions, or dental procedures, are routinely reported in the literature and are carried out with good outcomes [12].

Conclusion

Very few cases of post traumatic lower limb amputation in hemophiliac patients have been reported in the global medical literature. Amputations as an entirety in such cases, though slightly commoner, are most likely secondary to infections or hemophilic pseudotumours.

Majority of post traumatic amputations are seen to have been done within a week of trauma, not many cases of amputation within 24-48 hours have been noted. Secondary intention is also not a commonly deployed mode of healing for such stumps, and outcomes in general, have been unsatisfactory.

This case represents a unique clinical management dilemma, and expedient management with meticulous maintenance of Factor VIII levels, have contributed to the outcome.

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