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Case Report

Hemipelvectomy after severe pelvic injury in Factor VII deficiency toddler

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ABSTRACT

Traumatic hemipelvectomy is a lethal catastrophic injury. The reported average age of individuals surviving this trauma is 21 years old, suggesting the necessity of good physiological reserves to survive this type of injury. Dealing with this injury in children may call for special requirements throughout all the stages of diagnosis, treatment and rehabilitation. Experience in the resuscitation and subsequent treatment of individuals suffering from this traumatic condition in the paediatric population is even scarce. There are only several reported cases involving children and none of the paediatric cases suffered from comorbidities prior to their traumatic injury. The present report describes the successful management of a 16-month-old child with a medical history of a rare bleeding disorder a severe coagulation Factor VII deficiency who underwent right-sided traumatic hemipelvectomy.

Introduction

Traumatic hemipelvectomy is a lethal catastrophic injury, which is defined as an unstable ligamentous or osseous hemipelvic injury with rupture of the pelvic neurovascular bundle [1]. The common mechanism is high-energy transfer trauma [2], which is associated with a high percentage of death and morbidity [3]. This injury is quite a rare entity, with an estimated incidence of 0.6–2.4% of all pelvic fractures [1,3–5]. The exact number is unknown, probably because some of the victims die before reaching the hospital [6]. The determinants of survival appear to be related to rapid and effective vascular control, presence of other injuries, the quality of resuscitation, the time between injury and arrival at the hospital, the timing and quality of surgical intervention, and the presence of pelvic sepsis [3,7–9].

Dealing with this injury in children (below 13 years old) may call for special requirements throughout all the stages of diagnosis, treatment, and rehabilitation. There are only several reported cases involving children [1–3,10–16], with the first reported case by Lipkowitz et al. [16], who presented the successful management of a 7-year-old boy [15]. In a recent review of 52 cases presenting with traumatic hemipelvectomy or hindquarter amputation, only four were children [3]. Calonge et al. [13] reported the survival of two paediatric patients, with one 18-month-old girl the youngest child as of yet [12]. It should be noted that none of the paediatric cases suffered from comorbidities prior to their traumatic injury.

In the current case study, we present a case of traumatic hemipelvectomy of a very young child with a rare coagulation disorder of Factor VII deficiency. An extensive search of the literature did not reveal any such reported cases. Accordingly, we

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believe that adding this case to the current pool of experience will provide guidance to other clinicians in the management of this invasive and destructive trauma in children with a serious pre-comorbidity.

Case report

A 16-month-old boy was admitted to an acute care hospital half hour after being run over by a truck over his pelvic region during a motor vehicle accident. The child was transferred to the hospital via an ICU ambulance, and the emergency team called in the case in advance. The paediatric haematologist, who was familiar with the child's medical history of severe Factor VII deficiency and his routine medication of recombinant activated Factor VII (rFVIIa), was among the team awaiting the child at the emergency room upon arrival approximately one hour post-injury. The child arrived in severe haemorrhagic shock, GCS (Glasgow Coma Scale) 13, with a pulse rate of 220 beats per minute, blood pressure of 70/30 mmHg, respiration rate of 40 breaths per minute and transcutaneous oxygen saturation of 80% (room air). Upon arrival, he received one dose of rFVIIa, and then was sedated and intubated, immediately followed by urgent volume resuscitation, including packed red blood cells. The fluid resuscitation was done via his Port-A-Cath that was already in place.

The physical examination revealed the following: Pupils – PERLLA; no signs of head, chest, or abdominal injury; FAST (Focused Abdominal Sonography for Trauma) examination – negative. Clinical pelvic examination demonstrated rotational/vertical instability of the pelvic region, reduced capillary refill bilaterally, and no pulses in the right lower limb. A large, 12-cm-long, open wound exposing the right iliac bone was present in the right groin and thigh region. The wound was bleeding actively, and the right side of the scrotum was lacerated. In addition, an extensive crush injury to the left thigh was noted. The rectal examination showed no damage or bleeding, and the anal sphincter was intact.

The medical history of Factor VII deficiency was confirmed from the medical chart at the hospital. Factor VII deficiency was diagnosed at the age of 2 months due to easy bruising and positive family history for the disease. On the molecular level, the patient was found to have homozygous T359M mutation with R353Q polymorphism at the gene for coagulation Factor VII on chromosome 13q34. At the age of 4 months, spontaneous intracranial bleeding complicated by hydrocephalus had been diagnosed, and a ventriculo-peritoneal shunt was inserted. The patient was put on a prophylactic regimen of rFVIIa, with a dose of 30 μg/kg administered three times a week via a Port-A-Cath. After completing one year of secondary prevention, the regimen was changed to preemptive therapy, which entailed infusing the factor after minor trauma without removing the permanent central line. The present car accident occurred only a few days after switching to this protocol.

An hour and a half after admission resuscitation and improvement in the child's haemodynamic status, angiography was performed due to the absence of pulses in the right lower limb and the suspicion of active pelvic bleeding. The angiography demonstrated an avulsion of the right hemi-pelvis and a rupture of the right internal iliac artery (Fig. 1), which was treated by angiographic embolisation.

The child was transferred to the operating room (two and half hours after the admission) for an urgent surgical intervention by both a vascular and an orthopaedic surgeon. Exploration of the open pelvic wound revealed a torn right iliac vein below its junction with the vena cava. In addition, the right iliac artery was also ruptured at the point of its division into external and internal branches. Ligation of the divided vessels was performed in order to restore homeostasis, followed by a right ileo-femoral bypass using a vein graft. Finally, a repair of the skin laceration on the right scrotum was performed. After surgery (5 h after admission to the hospital), the child was hemodynamically stable and was transferred to the Paediatric Intensive Care Unit. Severe metabolic acidosis was evident upon arrival to the intensive care unit, though a clear and fast response was observed with ongoing fluid resuscitation.

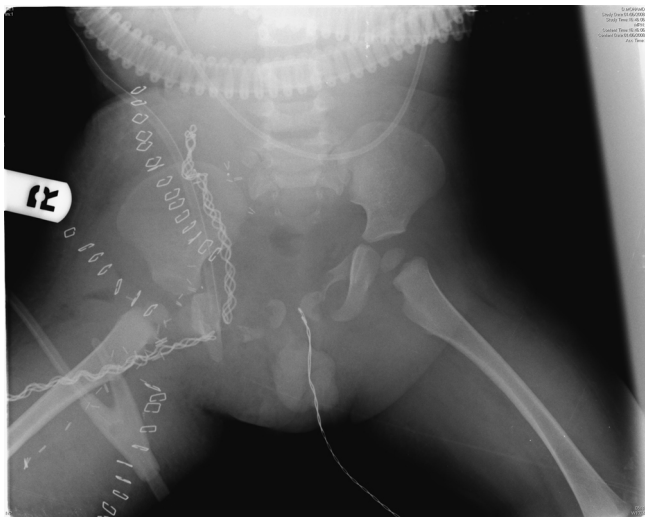

Fig. 1. Radiology after initial debridement and vascular restoration, showing disruption of right sacro-iliac joint and comminuted fracture of both Rami Pubis with displacement of the hemipelvis.

On the first postoperative day, the temperature of the right lower limb was lower in comparison to the contralateral side; however, the capillary refill was preserved. On the second postoperative day, ischaemic changes appeared in the right lower limb, with signs of compartment syndrome. The child was returned to surgery on the second postoperative day for a fasciotomy and revision of the vascular graft. During the surgery, an occlusion of the vascular graft by a thrombus was identified and opened by embolectomy. Following these procedures, the acid-base balance was normalised, and the child was extubated on the sixth day after the surgery.

Even following the vascular revision, there were still clinical findings of swelling in the right lower limb, absence of pulses, and prolonged capillary refill. The ischaemia of the right lower limb progressed, with development of a local and systemic infection caused by *Serratia marcescens*. Physical findings of extensive necrotic wound appeared in the right iliac region.

Despite treatment with local debridement and broad-spectrum antimicrobials (carbapenems), the septic condition progressed and a gangrene of the entire right lower limb developed (Fig. 2). The wound in the right iliac region became wider, with the appearance of uncovered Crista Iliaca bone. Additionally, skin necrosis developed on the lateral side of the upper third of the left thigh.

On the 12th day post-admission, a multidisciplinary consultation meeting was called due to the patient's life-threatening septic condition. The team included paediatric orthopaedic surgeons, a paediatric surgeon, a vascular surgeon, a plastic surgeon, a paediatric haematologist, a specialist in paediatric infectious diseases, and a PICU specialist. It was finally decided that due to the life-threatening situation, a hemipelvectomy must be performed despite the high risks of the procedure, particularly in light of the young age of the child and his coagulation deficits.

That same day, the child was operated on by a senior team, which included a paediatric orthopaedic surgeon, a vascular