Claw Toes Correction and Factor XIII Deficiency – A Case Report

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Summary

A 26-year-old male presented with claw toes and Factor XIII deficiency. Correction for his deformity was undertaken. Pre, intra and post-operative transfusions of plasma and blood prevented any haemorrhagic complications.

Key Words: Claw toes, Factor XIII deficiency

Introduction

Orthopaedic surgery is now practical and commonplace in conditions with abnormal blood coagulation such as haemophilia. The pre, intra and post operative regimes are standardised and though the surgical risks are higher in this patient group, even major operations such as joint replacements can be performed successfully. Surgical management in rarer coagulation disorders is not as well standardised and indeed these disorders are often unrecognised until during or after the operation with occasionally disastrous results.

Factor XIII (Fibrin Stabilizing Factor) deficiency was first described in 1960. Factor XIII is present in both plasma and platelets and is converted to an active form XIIIa by thrombin with the help of calcium. Factor XIIIa stabilizes the clot by chemical and mechanical bonding of adjacent fibrin molecules. Stabilized fibrin is not susceptible to lysis and produces effective haemostasis. These patients also exhibit defective wound healing. A diagnosis is based on the level of factor XIII in the plasma being less than 1% of normal[1].

We would like to report a patient who required an operation for correction of claw toes and who had been diagnosed to have this rare haematological condition.

Case Report

M. N., a 26-year-old male was referred to Orthopaedic Surgery with toe deformities in the right foot and difficulty in wearing shoes. The patient had been diagnosed with Factor XIII deficiency in 1981 after an initial misdiagnosis of Factor VII deficiency. He works as a forklift operator and since diagnosis has led a fairly normal life. He is the product of a consanguinous marriage but there is no family history of bleeding disorders. He has had recurrent bleeds in the right calf prior to diagnosis.

Examination of the leg showed wasting of the calf and equinus deformity of the right foot. Passive dorsiflexion of the foot was possible to plantigrade position only and increased the clawing of the 2nd and 3rd toes. In plantarflexion the 2nd toe could be straightened but the 3rd toe was not correctible (Fig. 1).

He was admitted for correction of the toe deformities.

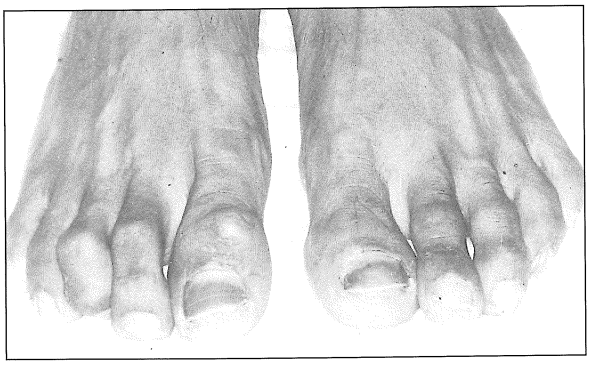

Fig. 1: Appearance of toes at presentation

He received a transfusion of three units of plasma on the day prior to the operation under intravenous hydrocortisone 100mg and promethazine 25 mg cover as he has manifested a skin rash with previous transfusions. He underwent surgery (Flexor to extensor tendon transfer and longitudinal Kirschner wire fixation) on the 2nd and 3rd toes. Intra-operatively, he was transfused 1 unit of whole blood. Since the Kirschner wires would remain in situ for 3-4 weeks, one unit of plasma was transfused as a booster 2 weeks after surgery to ensure effective coagulation for the entire period. Stitches were removed at 2 weeks and the Kirschner wires at 3 weeks after surgery.

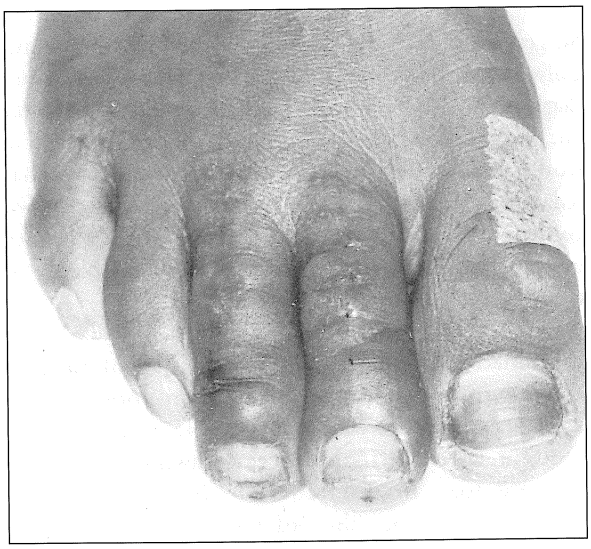

Fig. 2: Appearance at six weeks after surgery

Correction of the toes was well maintained and there was no problem with haemorrhage at any time (Fig. 2).

Discussion

Factor XIII deficiency is inherited as an autosomal recessive trait. Parental consanguinity is common. In most patients the disorder manifests shortly after birth or in childhood. Bleeding may be delayed for 12 to 36 hours after trauma although it can be immediate[1]. Acquired deficiency can occur in liver disease, secondary to disseminated intra-vascular coagulation or in the presence of inhibitor to Factor XIII. Intra-cranial bleeds are associated with a high mortality rate (30%).

The two important features of this disorder are as follows. Blood coagulation tests are normal except those for fibrin stabilization. All routine tests such as prothrombin time, partial thromboplastin time, thrombin time and bleeding time are normal. Platelet count is normal in heterozygotes. The clot is soluble in 5M urea or acetic acid in these patients. Quantitative assay for factor XIIIa is available. Secondly, bleeding may manifest only at one or two days after surgery. After minor operations or day care surgery where the deficiency has not been suspected and patient discharged, bleeding can occur away from hospital and endanger the patient's life.

Treatment for factor XIIIa deficiency is simple and effective. Less than five per cent of factor XIII are required for hemostasis and its half life is above twelve days in vivo. Two to three ml of plasma/kg body weight can provide adequate levels up to four weeks. Cryoprecipitate contains one and half to four times the factor XIII as compared to plasma[2]. Prophylactic infusion of five hundred ml of fresh frozen plasma or four to six bags of cryoprecipitate every three to four weeks is sufficient to attain therapeutic levels of factor XIII and surgery more complicated than the one this patient had can be safely performed[1]. For an actively bleeding patient five hundred ml of plasma every two days will suffice.

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Non-fatal Strangulation : An Uncommon Parachute-related Accident

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Summary

A case of non-fatal strangulation of the neck by rigging lines of a parachute during military training is presented. It is an unsual but potentially life-threatening injury. Probable factors leading to such injury are discussed.

Key Words: Parachute, Accident, Strangulation

Introduction

Parachuting, once regarded as a mean of transporting troops to battle fields, has now become recreational. With the formation of many amateur and professional parachuting clubs, increasing number of people all over the world can now enjoy the excitement and challenge of parachuting. Although adventurous, it is not without risks or dangers. This case report describes an unusual but potentially life-threatening injury which occurred in service parachute trainee in Kubang Kerian, Kelantan.

Case Report

A 22-year-old Royal Malaysian Air Force female trainee, made her maiden jump from an altitude of about 10,000 feet while undergoing service parachute training in Kelantan, Malaysia. During the initial free drop from the plane, she suddenly realised that one of the parachute's rigging lines was tied around her neck. She felt strangulated and lost consciousness momentarily, but, somehow managed to free herself from the entanglement within approximately 4 seconds and landed safely after a further 6 seconds.

On arrival at the Accident and Emergency Unit of Hospital Universiti Sains Malaysia, she was fully conscious and well orientated. Her only complaint was a severe pain in the neck with some difficulty in swallowing. Neck examination revealed the rope mark extending from the midline posteriorly to the left side of the neck, crossing the anterior neck and ending on the right side of the neck. A uniformly constricting band of about 5cm width was produced with areas of