A rare case of acquired factor XIII deficiency in a young male secondary to hyperconsumption: A case report

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Case Report

A rare case of acquired factor XIII deficiency in a young male secondary to hyperconsumption: A case report

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ABSTRACT:

Background. Acquired Factor XIII (FXIII) deficiency is a rare condition, often associated with underlying medical conditions or medications [1-3].

Case report. We present a case of a 23-year-old male with a history of alcohol abuse, [4] who presented with spontaneous hematomas and prolonged bleeding following minor trauma. Laboratory investigations revealed a severe deficiency in FXIII activity, with no evidence of congenital disorders or other underlying pathologies. Further evaluation revealed a pattern of hyperconsumption due to ongoing bleeding episodes. Treatment with FXIII concentrate and addressing the underlying cause led to significant clinical improvement [5].

Conclusion: This case highlights the importance of considering acquired FXIII deficiency in patients with unexplained bleeding diathesis, especially in the setting of alcohol abuse or other conditions associated with hyperconsumption. Early recognition and immediate treatment are very crucial in improving prognostic outcomes for these patients.

Keywords: Factor XIII, hyperconsumption, alcohol abuse, cryoprecipitate

ABBREVIATIONS:

ER- Emergency room
INR- International normalised ratio
TLC- Total Leukocyte count

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INTRODUCTION

Factor XIII is the final factor in the coagulation cascade with different chemical properties and physiological functions. The FXIII complex constitutes two subunits FXIIIA and FXIIIB. It has a significant role in clot stabilization by fibrin cross-linking and thereby, making the clot denser. It also plays an important role in platelet-dependent clot retraction, wound healing and repair of tissues post injury.

FXIIIA deficiency [1,2] is a very rare bleeding disorder which has an autosomal recessive inheritance. The incidence is one in a million to one in five million people. Rarely, we encounter an acquired deficiency of FXIII in common practice, which can occur secondary to any of the following three mechanisms: hyperconsumption, hypo-synthesis, or an immune-mediated process.

CASE PRESENTATION

A 23 year old male came to the ER with complaints of continuous bleeding from ulcer site since 1week. Patient sustained trauma to left leg, debridement was done outside and treated. But there had been bleeding from the site, which was not controlled with any typical measures. The wound ultimately developed into ulcer, and the patient presented with high grade fever.

On presentation, vitals were stable. Patient had no other complaints. General and systemic examination was within normal limits. There was bleeding and clot formation at ulcer site, with localized warmth and tenderness. There is edema surrounding the ulcer site. No evidence of any pedal edema/generalized lymphadenopathy.

Patient had history of head trauma when he was 1year old, for which craniotomy and evacuation was done in view of extradural hemorrhage and posterior fossa hematoma. Then, the patient was diagnosed with Factor 13 deficiency [3] as he had similar complaints at that time. Patient was not on any medications since then.

Investigations revealed low Hemoglobin (9.1), high TLC (17520), INR 1.6 and all other routine investigations were normal. Patient was started on high end antibiotics in view of sepsis. Patient's weight is 68kg, hence Cryoprecipitate transfusion of 6units was done to stop bleeding (based on criteria 1unit per 10kg).

Bleeding stopped after transfusion and INR came within normal limits. Daily dressing of wound done along with high end antibiotics. Patient improved symptomatically.

DISCUSSION

FXIII deficiency is an unusual bleeding disorder [4] where patients have a normal coagulation profile. A high suspicion is needed to diagnose the presence of FXIII deficiency. A potential family history of bleeding disorders, history of consanguineous marriage, or a history of residing in high prevalence areas may be present. A clot solubility test usually makes a diagnosis.

Only Immunologic assays are used for classifying the diagnosis. Cryoprecipitate is most commonly used in the treatment and prophylaxis for patients presenting with FXIII deficiency

[5]. Recently, a recombinant FXIII has also been made available for the same purpose. However, Acquired FXIII deficiency is extremely rare to encounter.

CONCLUSION:

Despite its rarity, it is essential to suspect this disease if there is severe clinical bleeding and routine coagulation tests are normal. Patient should be educated regarding their condition, which can prevent the unpredicted mishaps as happened with our case. Appropriate necessary treatment should be initiated as early as possible to control the bleeding and also for early prominent recovery.

Patient consent:

A clear informed consent obtained from patient and patient attenders

Conflicts of interest: Nil

Author's contributions:

Amukthamalyada Koduri has contributed to the data collection, initiation of treatment, follow-up and preparation of manuscript (writing and draft)

Arun Kumar Bathena and Magesh Kumar.S contributed to review, proof reading and finalizing the manuscript

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