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Case Report

Treatment Plan And Dosage For Acquired Fx Deficiency With Factor X Concentrate.

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Abstract

A uncommon disorder known as acquired factor X (FX) deficiency can result in potentially fatal hemorrhage. Here, we provide a human FX concentrate-based effective gastrointestinal bleeding (GI) management approach. Description of the case: A 61-year-old A male patient had a prolonged prothrombin time and upper gastrointestinal hemorrhage. An acquired FX deficit was found to be subsequent to AL amyloidosis, according to investigations.Results: Bleeding symptoms were successfully stopped by treatment with FX concentrate to maintain trough FX levels >20%, and levels >50% made urgent invasive treatments possible. Conclusions: This case study offers important new information about how to treat this uncommon ailment and how to use FX concentrates in acquired FX insufficiency.

Keywords : *Coagadex, amyloidosis, and factor X.*

INTRODUCTION

A rare hereditary condition called factor X (FX) deficiency causes a variety of bleeding symptoms, ranging from mucosal bleeding to potentially fatal hemorrhages [1–3]. Primary amyloidosis (AL) is the most frequent cause of the acquired variant, with aberrant proteins that are insoluble as a result of an enlarged plasma cell clone's excessive light chain release [4]. 8.7–14% of AL amyloid had an FX deficit [3, 5]. A substantial decrease in half-life and a quantitative deficit result from circulating FX being adsorbed onto amyloid fibrils.The management of these cases is not well supported by evidence. In the past, prothrombin complex concentrate (PCC), recombinant activated factor VII (rVIIa), and fresh frozen plasma (FFP) were used for factor replacement [6];There have been reports of thrombotic events and fluid overload problems, and therapy is frequently ineffective because FX is quickly removed from the bloodstream. In recent years, the emergence ofFor hereditary FX insufficiency, FX concentrates are used. Only three examples of acquired FX insufficiency have been documented in the literature, indicating the extremely limited experience of such concentrates in this condition [7, 8].Here, we show how FX concentrate can be used to treat bleeding in acquired FX deficiency and provide an example of a successful once-daily dosage schedule.

CASE DETAILS

Patient Information

A 61-year-old man arrived with melaena and excruciating stomach ache. He reported experiencing weight loss, ankle edema, and macroglossia for six weeks. Barrett's oesophagus was discovered during a recent gastroscopy for recurrent reflux, but no other noteworthy comorbidities were found. Neither a family history nor frequent drugs were present.

Clinical Findings and Diagnostic Assessment

Investigations and the initial symptoms pointed to a large upper gastrointestinal hemorrhage. An OGD was scheduled, but without anticoagulation, the INR was 2.9. The patient then experienced hemodynamic instability as his hemoglobin fell to 83 g/L (the recommended range is 133–167 g/L). With a normal activated partial thromboplastin time (aPTT) of 29.7 s (range 29–30 s) and fibrinogen of 4.58 g/L (range 1.5–3.5 g/L), the prothrombin time (PT) was elevated at 36.9 s (range 9–13 s). Coagulation throughout history Screens were standard.

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According to mixing studies, the PT was corrected to 15.0 s, and incubation had no effect. The measured FX activity was 7.0% (range: 50–150%). Using a modified prothrombin time assay on the ACLTOP coagulation analyzer, the FX assay was carried out.After adding FX-deficient plasma to diluted patient plasma, the PT was assessed. The observed PT, which is proportional to the FX activity, was used to compute the FX concentration (% activity), which was then plotted against a calibration curve. Additional factor activity tests comprised factors V (173%, range [50–150%]), VII (115%, [range 50–150%]), and II (92%, [range 50–150%]).

Therapeutic Intervention

There was a suspicion of amyloidosis and acquired FX deficiency. First, PT (23.8 s) and FX improved after receiving 10 mg IV vitamin K and prothrombin complex concentrate (Beriplex, 4000 units, CSL Behring, Marburg, Germany). (18%), although the symptoms of bleeding did not significantly change. The patient was taken immediately to a comprehensive care center for hemophilia.In accordance with the dosage for hereditary FX deficit (body weight × target FX rise × 0.5), Coagadex (human coagulation FX, Bio Products Laboratory Limited, Borehamwood, UK) was started once daily. 30-minute post-dose values obtained following the injection of FX concentrate showed some reduction in bleeding and an FX level of 24.0%.

However, successive dosages were doubled (determined by body weight x desired FX rise) in order to establish hemostasis, resulting in a post-dose FX of 36.7%. Adrenaline and hemostatic clips were used after an emergency gastroscopy revealed severe diffuse oesophageal hemorrhage with a friable mucosa. FX concentrate was dosed to achieve FX of >50% in order to simplify this procedure, and no substantial post-procedural bleeding was found.

Follow-Up and Outcomes

With the exception of modest but continuous bleeding from chronic hemorrhoids, the patient's symptoms stayed constant over the next two weeks. The bleeding gradually stopped after this was carefully treated with daily FX concentrate. Thrombotic symptoms were absent.issues that arise throughout therapy.BNP of 26,409 mg/L, urine albumin–creatinine ratio of 10.4 mg/mmol, and a kappa:lambda light chain ratio of 484 [kappa light chain 2372 mg/L] all corroborated the diagnosis of AL amyloidosis with gastric, renal, and cardiac involvement. Although 9% of plasma cells were identified by bone marrow aspirate, the trephine sample was insufficient. To prevent recurrent intrusiveFurthermore, the samples from the previous gastroscopy were examined again and submitted for Congo red staining, which showed traditional apple-green birefringence in polarized light (Figure 2).Velcade, thalidomide, and dexamethasone chemotherapy were started for the patient, but they were put on palliative care soon after since they could not handle the treatment.

DISCUSSION

A unusual consequence of amyloid fibril deposition brought on by an underlying plasma cell tumor is acquired FX insufficiency [3]. Bleeding symptoms usually manifest below FX levels of 10% in inherited diseases, whereas the threshold for bleeding in acquired diseases occursreduced, with one study showing a higher risk of bleeding below 25% [3].Other reasons, such as tiny vessel fragility from amyloid infiltration, deficiencies in other coagulation factors, abnormal fibrin polymerization, vitamin K deficiency, and thrombocytopenia/dysfunctional platelets, could be the origin of these hemostatic problems in AL amyloid [4]. The adsorption of FX by amyloid deposits is thought to be the main underlying mechanism.Furthermore, FX is bound by the macrophage scavenger receptor class A member 1 (SR-A1), which causes it to be internalized and degraded [9]. Pentaxin-2 opposes this action by forming a complex with FX/SR-A1 that stops internalization. Amyloid Fibroids may cause pentaxin-2 levels to drop, which would accelerate FX depletion [9].Treatments with FFP, PCC, and rVIIa are linked to thrombotic side effects and fluid overload, which is especially troublesome in AL amyloid patients who are weak and have heart problems. The management of inherited FX insufficiency has changed due to the availability of high-purity FX concentrates; nevertheless, little is known about their application in acquired deficiency.A single-factor concentrate called Coagadex has a license to treat hereditary FX deficiency. Here, we show how FX concentrate can be used to efficiently manage bleeding symptoms and make invasive treatments easier. However, the literature indicates We show that trough levels >20% resulted in the end of bleeding symptoms, despite the fact that FX < 25% increases the risk of bleeding. Additionally, despite the substantial cardiac amyloid burden, the patient did not experience pulmonary oedema or thrombotic problems. Although it took more frequent dosage to accomplish this, one report (n = 2) showed successful bleeding control with levels >15% [7].In this instance, we were able to maintain trough levels above the intended threshold of 20% by administering a once-daily dosing regimen after raising the dosage of FX concentrate by 100%. The differences between The variety of amyloid deposition and its subsequent effects on factor levels may be reflected in these examples. Here, FX concentrate was used to successfully control significant bleeding symptoms.In order to prevent bleeding issues, bleeding symptoms were managed by keeping FX levels at 20% and facilitating invasive procedures at levels above 50%. It is noteworthy that considerably larger dosages of FX concentrate were needed than those advised.dosage schedule for FX insufficiency that is hereditary. In order to

prevent bleeding diatheses, especially in the vicinity of invasive procedures, there is a need for screening individuals with AL amyloid for FX deficiency. To identify the best FX concentrate management techniques, more information is required in this rare disease domain. We provide a customized strategy for FX concentrate dosage, threshold goals, and administration frequency.

Author Contributions

The manuscript and figures were written by A.R. and R.J.S. The author of the manuscript was L.G. The manuscript was written, examined critically, and supervised by C.F. Every author has read and approved the manuscript's published form.

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Institutional Review Board Statement

Ethical approval is not necessary because the manuscript is a case report, there was no experimental treatment, no randomization of participants, no protocol requiring any of the patients or service users involved to receive treatment, care, or services that deviate from accepted standards, and it was not intended to yield findings that could be applied to other situations.

Informed Consent Statement

The patient and their next of kin have given their documented informed consent for this paper to be published.

Data Availability Statement

The information is in the paper or can be obtained from the authors upon request.

Conflicts of Interest

Regarding the writing and/or publication of this work, the authors declare that they have no competing interests.

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