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RESEARCH ARTICLE

PERIOPERATIVE MANAGEMENT OF A PATIENT WITH INHERITED FACTOR VII DEFICIENCY AND CROHN DISEASE : A CASE REPORT AND REVIEW OF THE LITERATURE

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Abstract

Inherited factor VII deficiency is a rare coagulation disorder. There is no direct correlation between factor VII activity and the bleeding risk. We report the case of a 24 years old patient with Crohn's disease and inherited factor VII deficiency who underwent ileocecal resection. The patient was considered as a high bleeding risk and was thus substituted with factor VII perioperatively. In the presence of thromboembolic risk factors, pharmacological venous thromboembolism prophylaxis was also introduced.

Introduction:-

Inherited factor VII deficiency is a rare coagulation disorder. There is no direct correlation between factor VII activity and the bleeding risk. Perioperative management is based on expert opinion rather than recognized guidelines. We report the case of a 24 years old patient with Crohn's disease and inherited factor VII deficiency who underwent ileocecal resection.

Case report

A 24 years old patient admitted for the perioperative management of a Crohn's disease related intestinal stricture. The patient has a known inherited factor VII deficiency, and presented a bleeding requiring blood transfusion following a dental extraction when he was 8 years old. At 16 years old, the patient had a perianal fistulotomy under general anesthesia with perioperative factor VII substitution, and pathology revealed a Crohn's disease. The patient is since on Adalimumab and Mesalazine.

The current history was consistent with an intestinal occlusion secondary to an ileocecal valve stricture, presenting as a König syndrome that lasted for two months. Laparoscopic surgery was performed after the patient received 2milligrams (33mcg/kg) of activated recombinant factor VII (rFVIIa) intravenously, as preoperative coagulation studies showed a prothrombine time of 22,5 seconds (normal : 10 to13 seconds) with a normal activated partial thromboplastin time and a Factor VII activity of 27% . The surgery, an ileocecal resection with ileocolic anastomosis went uneventful with minimal bleeding. The patient subsequently received 2mg rFVIIa every 6 hours for five days until he was discharged, for a total dose of 666mcg/kg. Venous thromboembolic prophylaxis was administered for two weeks using subcutaneous enoxaparin 40mg.

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Discussion:-

Factor VII (FVII) is a serine protease zymogen synthetized by the liver exclusively. It is a vitamin K dependent coagulation factor. Its plasmatic concentration ranges from 0,35 to 0,6mg/l, which corresponds to the 70 to 140% rates commonly used by laboratories. Its half life is very short approximating 4 to 6 hours, and most of factor VII circulates as a single strand inactive form. As a unique feature, a minimal proportion of weakly active double strand Factor VII (1 to 3%) can circulate freely even in the absence of coagulation activation. The active enzymatic form is linked to Tissue factor (TF) which plays the role of a cofactor, increasing the catalytic activity of FVII. FVII becomes active through proteolytic cleavage by proteases such as activated factor X, activated factor IX and activated factor VII. The TF-FVII complex is considered as the initiator of coagulation in vivo, the process of coagulation being triggered by the binding of little amounts of free circulating activated FVII (FVIIa) to TF or resulting from the autoactivation of FVII inside the TF-FVII complex. The complex TF-FVIIa can then exert its enzymatic activity on factor IX and factor X, with the final product being the formation of a fibrin clot. This step is regulated negatively by the tissue factor pathway inhibitor (TFPI) [1]. The involvement of multiple factors explains the absence of correlation between the plasmatic values and the hemorrhagic risk in deficient patients.

Hereditary factor VII deficiency is rare with a prevalence of 1/500 000, and defining its clinical picture and treatment relies on data from international multicentric studies like the International Registry on Congenital Factor VII Deficiency (IRF7) [2] and the Seven Treatment Evaluation Registry (STER) [3]. This autonomic recessive disorder can be caused by various mutations and is thus heterogenous. The deficit can be quantitative (type I) or qualitative (type II). It can be completely asymptomatic in 30% of patients, even during surgeries, or it can present with severe to lethal bleeding ranging from epistaxis and ecchymosis to intracranial hemorrhage.

The diagnosis can be made during a family screening or in the context of bleeding. It is established in the presence of a prolonged prothrombin time and an isolated low FVII activity. The lower limit of the reference range of the FVII activity, defined as 70%, is based on the statistical analysis of a reference sample reflecting the normal population. However, data from the STER and IRF7 show that the deficit can be considered significant when values are lower than 26% [4]. Bleeding is uncommon for values exceeding 10%, and spontaneous hemorrhage is unusual when rates exceed 20% [2,5,6]. Data from the STER and IRF7 also showed that bleeding at presentation predicted subsequent bleeding risk [6]. It has thus been proposed to classify the bleeding risk using personal or family bleeding history and FVII activity. Patients with a high risk are those having a FVII activity lower than 2% and a bleeding history, while patients with a low risk have a FVII activity higher than 20% with no bleeding history [5]. When planning an invasive procedure, especially major, the bleeding risk cannot be underestimated, even in asymptomatic patients. These patients can bleed later in life [4]. Also, the bleeding anamnesis gives little information in younger children. Our patient, who was asymptomatic until the age of 8, bled after a dental extraction and needed a blood transfusion. As a consequence, we considered him as a high risk patient.

The ideal substitution for factor VII deficiency is recombinant FVIIa concentrate (rFVIIa). When unavailable, plasma derived concentrates, fresh frozen plasmas or prothrombin complex concentrate can be used. Practice is variable when it comes to the dosing and duration of rFVIIa substitution. For major surgery, a minimal dose of 13mcg/kg administered at least three times at 6 hours intervals was efficient in preventing bleeding [7]. For invasive procedures and minor surgery, a total cumulated dose of 20mcg/kg in one day was efficient in preventing bleeding [8]. It is suggested to continue the substitution until healing and hemostasis are guaranteed. If it is decided to proceed to surgery without substituting the patient, rFVIIa must be kept available.

FVII substitution can come with a risk of venous or arterial thrombosis, especially when other risk factors exist [9], like the presence of an inflammatory bowel disease. The use of antithrombotic prophylaxis in patients with factor VII deficiency is reported in the litterature. It has been suggested to give pharmacologic prophylaxis to asymptomatic patients with a FVII activity greater than 30%, or with a FVII between 10 and 30% with a history of thrombosis, and to avoid it in the presence of a severe deficiency [10].

Conclusion:-

As a conclusion, inherited factor VII deficiency is a rare coagulation disorder, and a bleeding risk exists in the presence of a hemorrhagic history or with a factor VII activity lower than 20%. The proposed substitution is a minimum dose of 13mcg/kg administered at least three times at 6 hours intervals for major surgery and a total

cumulated dose of 20mcg/kg in one day for minor surgery and invasive procedures. In the presence of thromboembolic risk factors, pharmacologic thromboembolic prophylaxis should be considered.

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