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Acquired Factor VII Deficiency Presenting as Cardiac Tamponade: A Case Report

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Abstract **Case Report**

FVII (factor VII) is vitamin K-dependently synthesized in the liver. Hepatopathies, vitamin K deficiency, or use of vitamin K antagonists are the causes of acquired deficiency. Other types of acquired FVII deficiencies are rare. However, based on literature the incidence might be underestimated. The clinical manifestation of acquired FVII deficiency varies greatly in severity; asymptomatic course as well as severe life-threatening bleeding diathesis and fatal bleedings have been described. In this case report, we discuss a unique presentation of a 79-year-old male who was found to have cardiac tamponade revealing a severe acquired factor VII deficiency. A discordance between a prolonged PT and a normal aPTT was found in the biology lab report. And the diagnosis was confirmed by obtaining a factor VII activity assay. His management involved correction of his factor VII deficiency with fresh frozen plasma and pericardiocentesis.

Keywords: cardiac tamponade, Acquired factor VII deficiency, pericardiocentesis, prothrombin levels, fresh frozen plasma

INTRODUCTION

Acquired bleeding conditions are often the result of deficiencies of several clotting factors. Acquired isolated defects of clotting factors are rare conditions. The most widely known is the factor X (FX) deficiency associated with amyloidosis. However, there are other conditions associated with clotting defects, which are often ignored or unrecognized such as factor VII (FVII) deficiency[1,2]. This is due to the fact that the defect may be only mild to moderate and be reflected only by a minor prolongation of the prothrombin time (PT). The correct diagnosis may be missed if the treating physician does not request specific coagulation factor assays to investigate the isolated prolongation of the PT.

In this case report, we discuss a unique presentation of a 79-year-old male who was found to have cardiac tamponade revealing a severe acquired factor VII deficiency.

CASE REPORT

A 79-year-old male, with no known chronic medical problems presented with a 15-days history of dyspnea on exertion and orthopnea, along with positional chest pain. He also endorsed a dry cough that had been ongoing for one month without hemoptysis. He denied fevers, chills, night sweats, unintentional weight loss, or recent trauma. He was not seen in a healthcare setting for years. He was not taking any medications at home and had no recent exposure to anticoagulants or antiplatelets. There was no family history of bleeding disorders. He endorsed some episodes of average abundance epistaxis during the previous months, but denied a personal history of bleeding from other mucosal surfaces such as gum bleeding, hematuria, hematochezia, melena, prolonged bleeding after dental procedures and a tooth extraction, swelling in joints, and spontaneous skin bruising. Physical exam was normal. Initial labs revealed a discordance in the coagulation profile, which is summarized in Table 1.

Table 1: Coagulation profile of the patient

Coagulation profile	
PT	16.2%
INR	4.14
aPTT	30 sec

On admission, a low prothrombin levels (PT) was noticed (16.1 %; normal range 70–100%); activated partial thromboplastin time (aPTT), and platelet count were normal. His haemoglobin concentration was 10.3 g/dL). Correction in vitro of PT following the addition of 50% normal plasma without incubation was observed.

Further laboratory workup of coagulopathy revealed a factor VII level of <6% (normal range 67-143%). the rest of his tested factor activity levels were found normal.

These findings confirmed the presence of an inhibitor to factor VII; the antibody titre was quantified as 1 Bethesda unit (BU).

A bedside ultrasound of the heart revealed a large circumferential pericardial effusion and right atrial collapse along with evidence of tamponade (Figure 1).

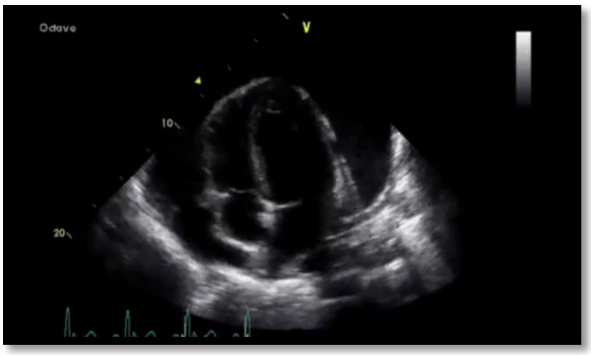

Figure 1: Massive pericardial effusion – transthoracic echocardiogram (TTE)

The patient was admitted into intensive care unit. Nonsteroidal anti-inflammatory drugs were not administered during his hospital course due to his high risk of bleeding. The initial plan to correct his factor VII deficiency in preparation for a pericardiocentesis was to administer 20 mL/kg of fresh frozen plasma (FFP) along with corticosteroid treatment. A pericardiocentesis with pericardial drain tube placement was performed on day 1 of admission with significant hemorrhagic drainage over the subsequent days.

Post-pericardiocentesis TTE revealed a residual, but stable pericardial effusion without evidence of tamponade. The decision to administer FFP during his hospital course was based on his continuous hemorrhagic drainage rather than a goal factor VII activity. Other options that were considered to reverse his coagulopathy were prothrombin complex concentrates (PCCs) and high-purity, plasma-derived factor VII concentrate (pdFVII). They were not readily available at our institution.

A thoraco-abdominopelvic computerized tomography (CT) scan was performed showed no signs of hematoma. The etiology of his tamponade was thought to be related to pericarditis, with coagulopathy due to factor VII deficiency leading to hemopericardium. The patient was discharged without active bleeding or reaccumulation of the pericardial effusion.

DISCUSSION

Acquired isolated factor VII (FVII) deficiency can be secondary to liver disease or vitamin K antagonists, and has been described in patients with malignancy, sepsis, post-operatively and in patients undergoing bone marrow transplantation.

It can also be caused by development of autoantibodies to this coagulation protein, which remains an extremely rare condition; actually only seven cases have been reported so far in the literature published in English [3].

The pathogenesis remain unclear. Kamikubo et al. demonstrated that the inhibitor had features of IgG1 with kappa and lambda-light chains inhibiting the procoagulant activity of activated FVII (FVIIa) by interaction with the light chain. They also suggested that the antibody recognizes the calcium-dependent conformation in or near the Gla domain having the ability to inhibit the interaction between FVIIa and tissue factor (TF) or phospholipid membranes [4]. In the case described by Weisdorf et al, an inhibitory antibody could not be identified but they reported an IgG autoantibody without inhibitory activity in a patient with aplastic anemia. They suggested that the antibody enhances in vivo clearance of FVII by formation of immune complexes [5].

From a clinical point of view. In contrast to the majority of blood coagulation factor deficiencies, FVII deficiency is characterized by a poor relationship between FVII clotting activity and bleeding tendency.

The patient described in our case report experienced a severe bleeding [6].

Acquired FVII deficiency is most often diagnosed in patients with discordance between a prolonged PT and a normal aPTT (figure 2). As a diagnostic step, coagulation factor activity should be determined an isolated decrease of FVII level should be confirmed [7-9]. In our case, the decrease was very significant <6%. To further investigate the FVII deficiency, mixing study should be performed. If the PT remains prolonged after mixing of patient plasma with

normal pooled plasma, a factor inhibitor is suspected [9].

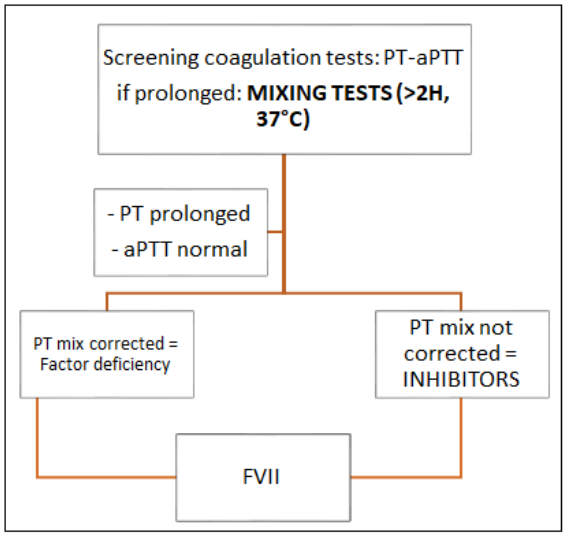

Figure 2: Identification of abnormalities of clotting factors

Currently, there are no standardised guidelines for the treatment of acquired FVII deficiency. The initial therapeutic choice must be made based on the biological features and clinical severity of bleeding [[i]].

We would suggest to start with plasma products in patients with low titre inhibitors (< 10 BU) and a moderate bleeding tendency. rFVIIa is the first choice treatment in cases with high titres of autoantibodies, severe (or even life-threatening) hemorrhage or lack of clinical response to plasma-derived products [10]. The addition of antifibrinolytic drugs may be interesting, especially in cases with mucosal bleeding (with the exception of haematuria) and they may occasionally be successful by themselves [11].

Thrombotic episodes have been reported in patients with and without inhibitors to FVII treated with high doses of FVII concentrates so they must be used with caution [12].

Besides, the underlying disorder that might be associated has to be treated. When antibodies are present and persist, immunosuppression is recommended to eradicate the inhibitor [13].

CONCLUSION

Acquired FVII deficiency is a rare disease, with only few patients reported in the literature so far, but the incidence might be underestimated. The exact pathogenesis of the disease is still unknown, but different pathophysiological hypotheses have been suggested. Bleeding occurs in only about the half of the cases. Patients without prior clinical evidence of bleeding are diagnosed based on coagulation abnormalities during routine blood screening. The overall complete remission is poor and bleeding complication are accounted for the death in a lot of patients. However, our patient was discharged without any further complication.

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