THROMBOSIS OF THE ABDOMINAL AORTA IN CONGENITAL AFIBRINOGENEMIA

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INTRODUCTION

Congenital afibrinogenemia is clinically characterized by a bleeding tendency. However, rare cases of thrombosis involving the arterial or the venous district have been described in patients with fibrinogen deficiency. Reported thrombotic events occurred either in association or not with replacement therapy or with pro-thrombotic risk factors.

The optimal antithrombotic therapy and its duration in these patients is unknown.

AIM

We describe an aortic thrombosis with peripheral embolism in a 48-years-old female with congenital afibrinogenemia.

CASE REPORT

The patient had a long-term bleeding history: hemorrhage from the umbilical cord 5 days after birth, bleeding of gums and after minor trauma. Congenital afibrinogenemia was diagnosed at 4 years of age. Hemoperitoneum due to ovarian cysts rupture occurred at the age of 16 years.

She was initially treated on demand with hemo-transfusions and cryoprecipitates, then with fibrinogen concentrates (FC). In 1990 HCV infection was diagnosed.

In 2001 she developed a spontaneous cerebral hemorrhage treated with FC, which were maintained as long-term prophylaxis (1 g i.v. every 10 days; Fibrinogeno TIM3, Baxter, replaced by Haemocomplettan P, CSL Behring, since 2004) without bleeding recurrences.

In april 2011 she was admitted for ischemia of the 4th right toe. She had reported an incidental fall with a low back trauma a few months before.

Arterial pulses and EchoColorDoppler of the right lower limb were normal, as well as an ECG and an Echocardiogram. An angio-TC of abdominal aorta showed a thrombosis from the origin of renal arteries to the carrefour with a stenosis of 50-60% of the lumen and a distal floating part of 34 mm (Figure 1).

Fibrinogen levels at admission were undetectable; PT and aPTT were uncoagulable. Mild hypercholesterolemia and arterial hypertension were seen, whereas vasculitis, thrombophilia, diabetes mellitus, retroperitoneal diseases, cancer, and atherosclerosis were excluded.

The patient received FC to maintain fibrinogen levels above 80 mg/dL; in addition, reduced dose of sc enoxaparin (75 U/Kg bid) and aspirin (50 mg every other day) were given as antithrombotic treatment.

The toe ischemia completely recovered. Antithrombotic treatment was maintained in addition to prophylactic FC (Haemocomplettan 2 g iv every 4 days); enoxaparin was replaced by fondaparinux 5 mg od on February 2012.

No bleeding complications nor thromboembolic recurrence were observed. After 12 months from the event aortic thrombus was reduced at an angio-TC control (Figure 3) .

FIGURES

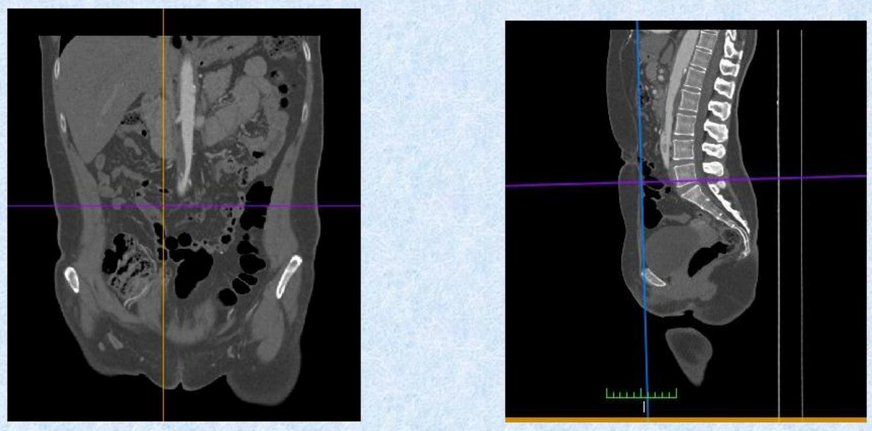


Figure 1: angio-TC of abdominal aorta at admission showing a thrombosis from the origin of renal arteries to the carrefour

Figure 2: Ultrasound scan of the abdominal aorta at admission confirming the presence of thrombosis

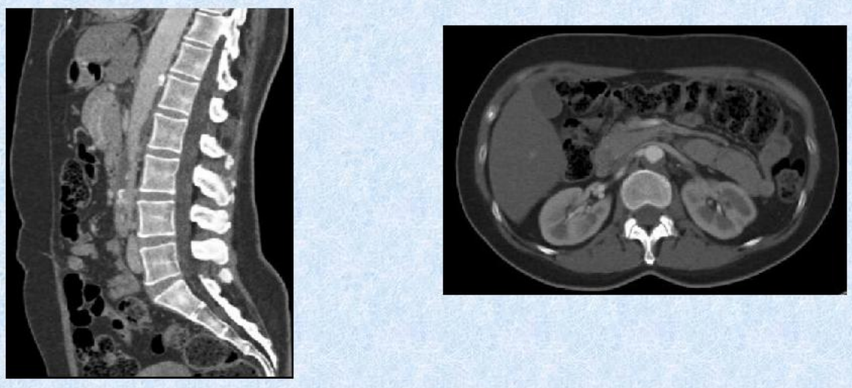
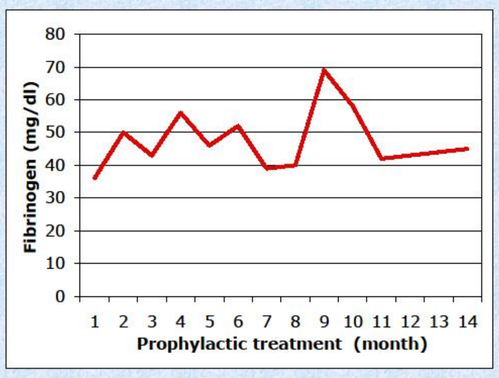


Figure 3: angio-TC of abdominal aorta after 1 year from the event showing a partial reduction of the thrombous

COAGULATION TEST DURING FOLLOW UP



Pre-infusion fibrinogen levels ranged between 40 and 70 mg/dl (figure).

PT values ranged between 37% and 50%, and aPTT values between 24 s. and 30 s. before fibrinogen administration.

CONCLUSIONS

We can speculate that thrombus formation in our patient might have been triggered by a traumatic lesion of aortic endothelium, since other thrombotic risk factors were ruled out. The role of fibrinogen prophylaxis remains uncertain.

Our therapeutic approach was empirical, but proved to be effective in obtaining thrombus reduction; in addition it was safe. Further studies are needed to optimize antithrombotic treatment in this peculiar setting.

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