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ABSTRACT N. PO07

TRAPIANTO DI FEGATO IN UN CASO DI AFIBRINOGENEMIA CONGENITA.

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Background: Fibrinogen (FBG) is involved in the final steps of coagulation as a precursor of fibrin monomers. Inherited FBG disorders are generally classified as quantitative or qualitative. Quantitative disorders include afibrinogenemia (aFBG), which is characterized by the complete absence of FBG. aFBG is the result of homozygosity or combined heterozygosity for the causative mutations. The diagnosis is based on the measurement of FBG activity and antigen levels which define the severity of the disorder. Clinical manifestations vary from being asymptomatic to developing catastrophic life-threatening bleeds or thromboembolic events. The main treatment for quantitative FBG disorders is FBG supplementation.

Case report: A 26-year-old patient with inherited aFBG, homozygous for nIVS7+1G>7 mutation, with immunologically undosable FBG, was on-demand treatment with human fibrinogen concentrate (HFC) for spontaneous bleeding until October 2022. Then he developed a porto-splenomesenteric (P-S) confluence thrombosis with intrahepatic extension complicated by intestinal ischemia that required segmental jejunal resection, mechanical thromboaspiration, packing of a Transjugular Intrahepatic Porto-systemic Shunt (TIPS) and percutaneous angioplasty. At postoperative re-evaluation CT scan found bilateral subsegmental pulmonary embolism, thrombosed TIPS and evolution of thrombosis to the entire PS venous axis, with subsequent cavernomatosis development. Low molecular weight heparin (LMWH)(8000Ux2 twice daily) was initiated, together with HFC (2g three times a week). On 30/01/24, in order to allow liver transplant (LT), a left spleno-renal shunt with autologous jugular vein was performed after infusion of HFC 4g 1h before the surgery. Monitoring of FBG was initiated in the first 48h postoperatively, with the need of an additional 4g of HFC to maintain FBG above 100 mg/dl. LMWH was started the +1 day at a dose of 6000U/day until preoperative prophylaxis was resumed on +8 day. On 15/11/24, after infusion of HFC 4g 1h before surgery, LT was performed with reno-portal transposition and additional 2g of HFC was needed the next day. LMWH 4000U/die was started on +1 day and continued in the following months to ensure liver perfusion and due to a severe adenovirus systemic infection. Normalization of FBG and no more need of HFC was achieved on +3 day (last assessments of 250 mg/dl).

Conclusion: We report one of the few cases of inherited aFBG, complicated with massive splancnic thrombosis successfully treated with LT.

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