

Letters to the Editor

Beyond Heyde's syndrome

Key words: Heyde syndrome. Multiple myeloma. Gastrointestinal bleeding.

Dear Editor,

The association of aortic stenosis and recurrent gastrointestinal bleeding due to angiodysplasia was first described by Edward G. Heyde in 1958 (1). This so called Heyde syndrome was recently associated with type 2A von Willebrand disease. According to this theory, when von Willebrand factor pass through the stenotic aortic valve, it is exposed to a high flow, which determined that its multimers are destroyed by a disintegrin (ADAMS 13). Such multimers are necessary to keep hemostasis in high flow conditions such as angiodysplastic arteriovenous malformations, frequently observed in the elderly (2). Aortic valve replacement has been proposed to treat this syndrome when a significant anemia is detected and medical and endoscopic treatment failed (3).

Case report

A 62-years-old female patient with a past history of double aortic disease (severe stenosis and moderate insufficiency) with preserved systolic function. The patient was admitted in another hospital two years before due to anemia and melena. Gastroscopy and colonoscopy were normal. Endoscopic capsule was performed and a bleeding spot was detected in the jejunum, followed by enteroscopy with sclerosis and marking of the two vascular lesions of the jejunum. CT scan was normal and tumor markers were negative. One year later, the patient was hospitalized in our unit due to the same symptoms. Gammagraphy with octreotide was

normal and an arteriography detected an arterial malformation in the second jejunal branch of the superior mesenteric artery. A new enteroscopy identified a new jejunal lesion distal to the previous ones which was coagulated with APC with successful hemostasis. A Heyde syndrome was suspected after cardiac failure, therefore aortic replacement with a biologic prosthesis was performed. Even though the patient did not have melena, slight leucopenia, trombopenia and anemia was detected in the blood count. A proteinogram was performed identifying slight monoclonal IgG gammopathy and Bence-Jones proteinuria.

Discussion

The prevalence of Heyde syndrome is unknown. The diagnosis has to be made ruling out other possibilities and requires the triad of aortic stenosis, acquired coagulopathy and anemia due to bleeding secondary to telangiectasias (intestinal or any other location) (4,5). Even though all three conditions were observed and the valve replacement was considered curative, her anemia did not improve. A more detailed study lead us to diagnosis of stage IA multiple myeloma. This study was not initially performed because we considered the anemia was secondary to gastrointestinal bleeding. We report this case considering an infrequent association of these two diseases and particularly because Heyde syndrome did not respond to treatment and enabled the diagnosis of another disease which caused anemia.

Alejandro Martínez-Caselles, Cristina Martínez-Pascual,
Antonio Sánchez-Torres and Fernando Carballo-Álvarez

*Department of Digestive Diseases. Hospital Universitario
Virgen de la Arrixaca. Murcia, Spain*

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