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ABSTRACT

This article includes the management of one clinical case. A 27 years old patient was urgently hospitalized with upper digestive hemorrhage diagnosis, caused by Dieulafoy disease. The primary diagnosis was confused and needs more pre- and intraoperative confirmations. Histological and morphological examinations confirmed the clinical diagnosis. The surgical treatment and intensive care contribute to removal of vital risk and better therapeutic outcome.

Key words: Dieulafoy disease; hemorrhage; histology, treatment

INTRODUCTION

Angiodysplasias of the gastro-intestinal tract are vascular abnormalities situated in the wall structure of digestive tract organs, from the esophagus to anus. They are often acquired and, more rarely, congenital, clinically appreciated or latent, evolving on their own or in association with other pathologies. Fontch P. defines angiodisplazia as an idiopathic syndrome consisting of ectasia of intestinal submucosa, with or without ectasia of overlying mucosa capillaries [3, 4, 6, 7, 8].

Dieulafoy disease represents an arterial angiodysplasia characterized by anomaly of vascular development of the stomach submucosa, with the presence of large arterial erosions in the absence of vasculitis with the formation of acute ulcers, often complicated with digestive haemorrhage [2, 4, 7]. Similar cases were described at autopsy by Gallard (1884). The detailed clinical picture was described by Dieulafoy G. (1998), being distinguished as a separate nosologic form – simple ulcers [3, 4, 5].

Clinical features of the disease are:

- It is characterized by proximal stomach lesion, localization of ulcers on the posterior wall, small curvature (60-80%), at a distance of up to 6 cm from the esophagus junction [2, 7];
- It occurs between 16-96 years (average 50-60 years), the ratio of men: women being 2:1;
- It is presented as a source of superior digestive hemorrhage (0.3-5.8%), recurrent (18-100%), cataclysmic hemorrhages in 1/3 of patients [1, 3, 4];
- Clinical development is characterized by: sudden onset without abdominal pain; active hemorrhage (hematemesis, melena) [2, 8];

Predisposing factors are: chronic alcoholism, salicylates and steroids administration, stress. The absence of ulcerative anamnesis is important [2, 8].

The Dieulafoy ulcer may be located on the way of the digestive tract and may be associated with other pathologies such as: gastric and duodenal ulcers, Mallory-Weiss syndrome, Crohn's disease, gastric cancer [2, 4, 6].

Histomorphologically it is characterized by: the presence of large arteries in the ulcerous defect of the stomach mucosa, dysplasia of muscular substrate vessels and lymphocytic infiltration [2, 4, 6].

Diagnosis of Dieulafoy disease in the multidisciplinary study of causes of digestive hemorrhages is based on endoscopic techniques (48-98%), selective angiography results (30%) and histomorphological study (20%). All of these methods are used to identify Dieulafoy disease, assess the localization of vascular fistula, and evaluate treatment outcomes [2, 4, 7].

Clinical case: Patient S., aged 27 years old, is hospitalized urgently for a specific symptom of superior digestive hemorrhage: repeated vomiting with fresh blood and clots, vertigo, headache, general weakness, melena. From the past, it is known that in 2012 he suffered the first upper gastrointestinal hemorrhage, and he was treated conservatively under stationary conditions. He was healed and discharged with the diagnosis of Mallory-Weiss syndrome, receiving recommendations for ambulatory treatment.

The general exam reveals that at the admission, the patient is in a general serious condition. The visible skin and mucosa are pale and clean. Cold sweats, hypotension – 90/60 mmHg, tachycardia, FCC-120 bpm /min.

The patient presented a hypovolemic shock. The nasogastric tube evokes gastric content –hematemesis, fresh clots. Rectal exam – rectal ampoule with changed blood content. Laboratory samples show post hemorrhagic anemia – Hb - 70 g / l; erythrocytes – 2.4 • 10^{12} g /l.

The admission diagnosis focused on Mallory-Weiss syndrome complicated with superior digestive hemorrhage gr. II-III; hypovolemic shock. The diagnosis has been endoscopically confirmed. Two flat defects of 1.8-1.3 cm and 1.5-0.3 cm, fibrin-coated, are determined in the esophageal junction. Emergency treatment in the intensive care unit includes correction of volume and hematic correction, antiulcer medication, hemostatic. The patient is in clinical and hemodynamic improvement. More than 18 hours after hospitalization, the hemorrhage is re-evaluated. Repeated endoscopic control is performed, the source of hemorrhage cannot be appreciated due to inconclusive endoscopic signs. The Blackmore probe for the Mallory-Weiss syndrome was applied, the upper digestive hemorrhage being stopped.

For a differential diagnosis of superior digestive hemorrhage with digestive angiodysplasia, repeated endoscopic control was performed over 48 hours after admission, attesting to the presence of 2 mucosal rupture of 2.0 cm and 1.4 cm in diameter, which covered the gastroesophageal junction with fibrin. In the posterior subcardial area on fornix – a 0.5 cm pitched defect with freshly fixed thrombus, laminar hemorrhage.

Endoscopic diagnosis: Dieulafoy's disease complicated with repeated, active digestive hemorrhage. Surgical treatment is indicated immediately.

Intraoperative endoscopic control confirmed: the stomach of normal size and shape, elastic walls. In the stomach lumen – coffee grounds hemorrhage, the glossy pale mucosa of the stomach. Preserved relief of the fornix, esophageal and cardiopulmonary varicose veins were absent. In the posterior subcardial area of fornix, a 0.5 cm pitched defect with a freshly fixed thrombus was assessed.

Gastric resection is practiced. In the proximal subcardial region of the stomach, on the large curvature - the posterior wall, at 3.0 cm from the esophagus junction, there is a mucosal ulceration of 3.0 mm, covered with a fresh thrombus, fixed without signs of active hemorrhage. Blood clotting in the jet was seen in the removal of the clot. The resection was performed in the direction of the large curvature (fig. 1). In the gastro-splenic ligament, large vessels were found which do not correspond to the anatomical realities.

The particularities of the clinical case: diagnostic and therapeutic problems with major impact on pre- and postoperative clinical development. The uncertain diagnosis has contributed to errors in therapeutic conduct. The histomorphological diagnosis confirmed the clinical diagnosis of Dieulafoy Disease (fig. 2, 3, 4).

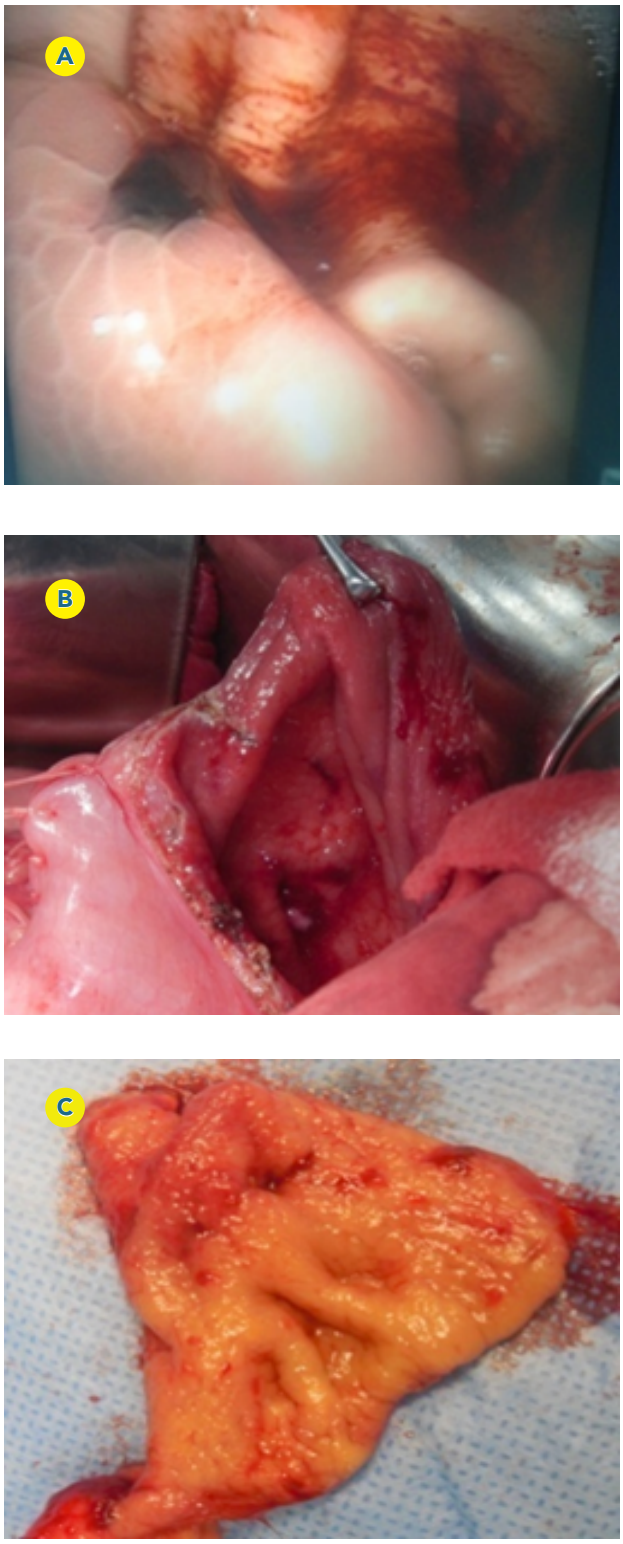

Fig. 1. a) Endoscopic view, b) Intraoperative, c) Postoperative

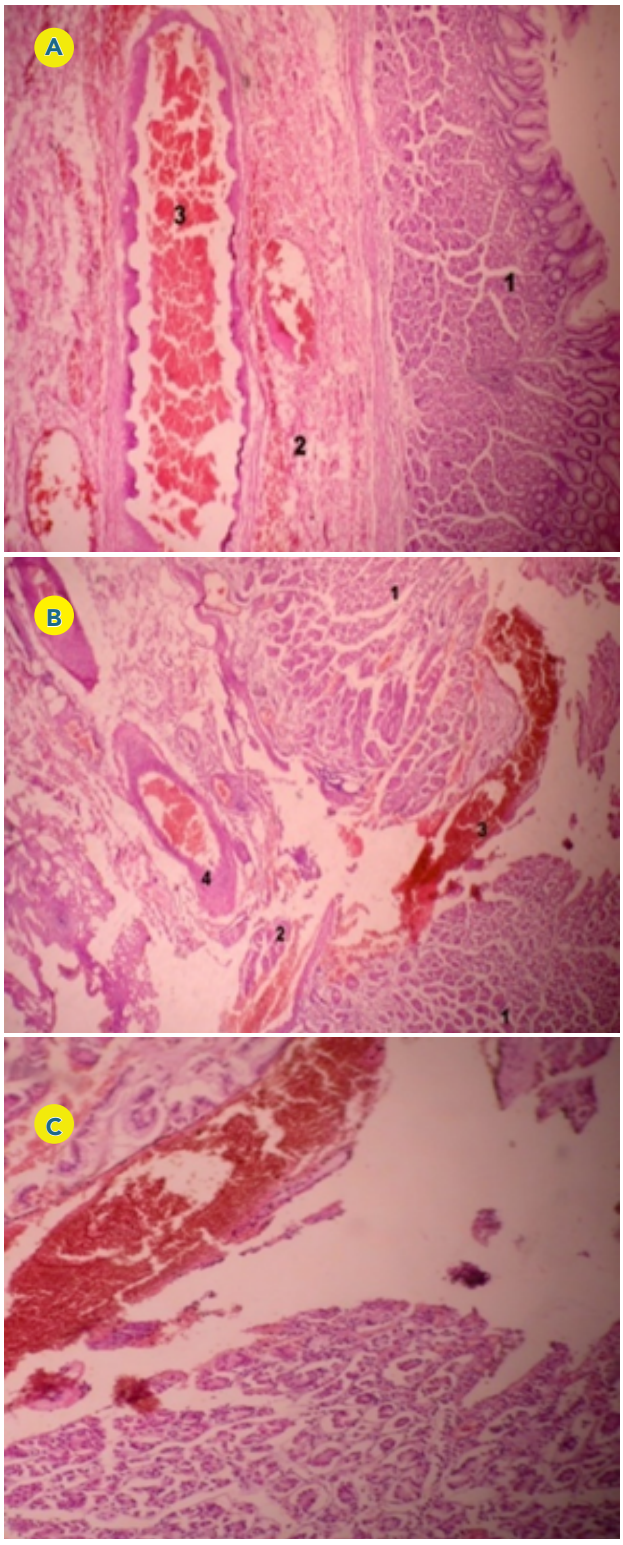

Fig. 2. A - piece of the gastric wall in the region adjacent to the Dieulafoy lesion: 1) mucosa without signs of inflammation, 2) sSubmucosa, 3) artery dilated with blood stasis. H-E. x100; B - A piece of the gastric wall in the region adjacent to the Dieulafoy lesion: 1) gastric mucosa without signs of inflammation, 2) artery with eroded walls, 3) hemorrhage and mucosal lesion adjacent to the eroded vessel, 4) Dilated arteries in submucosa. H-E. x40; C - hemorrhage outbreak with mucosal eruption. H-E. x200

DISCUSSIONS

Gastrointestinal angiodysplasias are responsible for approximately 6% of gastrointestinal tract hemorrhages and 1.2-8% of hemorrhages located in the upper digestive tract. This is the fourth cause of digestive hemorrhage after ulcer disease, varicose hemorrhage and colon diverticulosis, all of which cause 85% of digestive hemorrhage [1, 6, 7, 8].

Concomitant with the more frequent use of contemporary imaging methods (endoscopic and angiographic), the incidence of angiodysplasia diagnosis has increased, patients being included in the category of idiopathic digestive hemorrhage.

According to the Camillieri classification (1996), clinical, histological arterial-venous malformations are divided into [6, 8]:

- Type I – arteriovenous malformations with predominance of dysplastic veins with the small intestine / colon wall;
- Type II – Osler-Webber-Rendu disease;
- Type III – haemartomatous vascular lesions encountered in Peutz-Jeghers syndrome, blue rubber blue nevus and Klipper-Trenaunay syndrome;
- Type IV – gastrointestinal angiodysplasia, localized predominantly on the right colon, associated with aortic stenosis, (Heyde syndrome);
- Type V – Massive gastrointestinal bleeding caused by submucosa lesion with large vessel (Dieulafoy Disease).

CONCLUSIONS

Dieulafoy's disease is a rare cause of digestive bleeding, being identified as a cause of obscure bleeding. Diagnosis of pathology is difficult and complex (endoscopy, selective angiography and histological examination). EFGDS intraoperatively confirms clinical diagnosis. Surgical interventions are prevalent, and it is necessary to minimize "blind" resections.

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