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Hemorrhage from primitive rectal varices in patient with idiopathic thrombosis of portal vein: case report

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SUMMARY: Hemorrhage from primitive rectal varices in patient with idiopathic thrombosis of portal vein: case report.

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Introduction. Rectal varices, primitive or secondary to hypertensive or thrombotic disorders of mesenteric-portal circle, represent an uncommon cause of lower digestive bleeding. The presence of rectal varices associated to idiopathic venous portal thrombosis represents a distinct nosologic entity, with important clinical and therapeutic problems related to it.

Case report. Patient of young age, with positive anamnesis for primitive rectal varices, admitted to our department for a serious rectorragia. The laboratory underlined moderate anaemia and the endoscopy documented the presence of multiple rectal varices, without evident signs of bleeding; the endoscopy documented the presence of two esophageal small varicose cords F1. The hepatobiliary sonography and the portography showed the massive thrombosis of the portal vein. The new serious episode of rectal bleeding induced us to subject the patient to a surgical operation of Hartmann recto-sigmoid resection.

Conclusion. Because of the slight number of reported cases of primitive rectal varices and because of the scattering of many dates it's difficult to draw an univocal diagnostic and therapeutic algorithm. Clinical framing and subsequent therapeutic approach rise often up from personal experience rather than well defined guidelines. The treatment is controversial, time by time many therapeutic options are reported either conservative or interventist. The failure of conservative therapy and the recurrent episodes of bleeding give indication to surgical treatment, that is represented by Hartmann colonic resection and/or the porto-systemic shunts in the cases of portal hypertension; in our case we made colonic resection sec. because of lapsed performing status of the patient.

RIASSUNTO: Emorragia da varici primitive del retto in paziente con trombosi idiopatica della vena porta: caso clinico.

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Introduzione. Le varici rettali, primitive o secondarie ad una patologia ipertensiva o trombotica del circolo mesenterico-portale, rappresentano una causa non comune di emorragia digestiva bassa. La presenza di varici rettali associata a trombosi idiopatica portale rappresenta un'entità nosologica distinta, con importante problematiche cliniche e terapeutiche correlate.

Caso clinico. Un giovane paziente, con anamnesi positiva per varici rettali primitive, viene ricoverato presso il nostro reparto per un quadro di rectorragia severa. Gli esami di laboratorio dimostrano anemia moderata e l'endoscopia documenta la presenza di multiple varici rettali, senza segni evidenti di sanguinamento; l'endoscopia documenta la presenza di due piccole varicosità esofagee F1. L'ecografia epatica e la portografia mostrano una trombosi massiva della vena porta. Un nuovo e severo episodio di rectorragia ci obbliga a sottoporre il paziente ad intervento chirurgico di resezione retto-sigmoidea secondo Hartmann.

Conclusioni. A causa del numero esiguo di casi di varici rettali primitive riportati in letteratura e della dispersione di molti dati è difficile disegnare un algoritmo diagnostico terapeutico univoco; spesso l'inquadramento clinico e il susseguente comportamento terapeutico scaturiscono dall'esperienza personale piuttosto che da linee guida ben codificate. Il trattamento è controverso e di volta in volta vengono adottate opzioni terapeutiche conservative o interventistiche. Il fallimento della terapia conservativa e gli episodi ricorrenti di sanguinamento pongono l'indicazione al trattamento chirurgico che è rappresentato dalle resezioni coliche e/o dalle deviazioni porto-sistemiche nei casi di ipertensione portale. Nel nostro caso abbiamo fatto ricorso ad una resezione colica secondo Hartmann per lo scaduto performance status del paziente.

KEY WORDS: Primitive rectal varices - Idiopathic portal thrombosis - Lower digestive bleeding.
Varici rettali primitive - Trombosi portale idiopatica - Emorragia digestiva bassa.

Introduction

Rectal varices, primitive or secondary to hypertensive or thrombotic disorders of mesenteric-portal

circle, represent an uncommon cause of lower digestive bleeding (1, 2). The presence of rectal varices associated to idiopathic venous portal thrombosis represents a distinct nosologic entity, with important clinical and therapeutic problems related to it. In fact if the bleeding is at once an unusual manifestation, however it can be so severe, since the beginning, that it could be a risk for patient survival,

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especially in the cases of late diagnosis and unappropriate and untimely treatment.

We lately observed a case of primitive rectal varices associated with idiopathic portal thrombosis. In this case we put the attention on a clinical and diagnostic management and on the objective problems for the treatment of a rare pathologic condition that is not yet settled in a nosologic point of view and that is often severe in its evolution.

Case report

BA, male, 26 years old, affected by mioclonic epilepsy secondary to cranial trauma, hospitalized in our Operative Unit with diagnosis of rectal bleeding. About anamnestic data, the patient reported an endoscopic diagnosis of primitive rectal varices, made when he was 21 years, after a severe hemorrhagic event, that required urgent hospitalization and elastic ligation of numerous rectal vascular ectasias; instrumental exams made to complete the diagnostic iter (endoscopy, abdominal sono-tomography, portal color-Doppler) excluded other associated pathologic conditions. After this first event the patient had not more hemorrhagic events and was completely asymptomatic.

Income in our Unit the patient, affected by moderate anaemia (RBC 3.550.000, Hb 9.20), was subjected to a recto-sigmoidoscopy that proved, in the rectum until to recto-sigmoid junction, the presence of multiple varices of submucosal venous reticulum, with winding course, without obvious signs of bleeding (Fig. 1). An esophagogastrosocopy was performed, too that it showed a normal esophagus in its proximal and medium tract. Instead, in its lower third, the presence of two F1 blue little varicose cords was documented; cardia was gaping and gastric mucosa, especially in fundus and corpus, was widely turgid, hyperemic and overflowed by multiple petechial erosions. Bulbus and second duodenal parts were uninjured. Hepatobiliary sonography (US) showed a volumetric increase of the liver, with diffusely unhomogeneous echostructure and without focal lesions. US (color-Doppler) allowed us to prove a massive thrombosis of portal lumen, partly involving upper mesenteric vein and splenic venous axis, in correspondence of cephalic segment of pancreas,

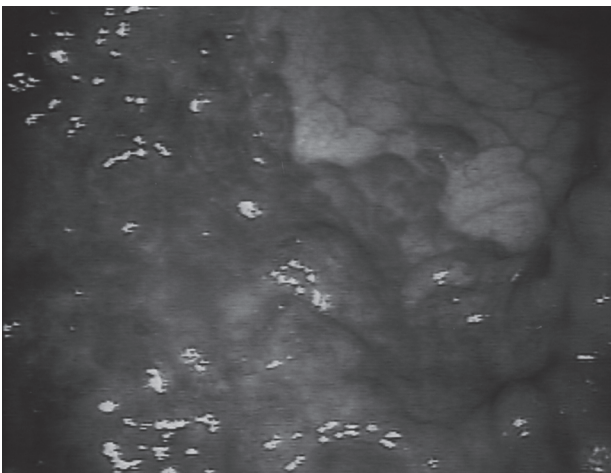


Fig. 1 - Rectosigmoidoscopy: presence in the rectum until to recto-sigmoid junction of multiple varices of submucosal venous reticulum, with winding course, without signs of bleeding.

with multiple ilar collateral vascular beds. This condition was confirmed by a following angiographic exam (portography).

In concurrence with a new hemorrhagic episode of moderate entity an endoscopic application of haemostatic loops in the venous ectasies in the rectum was performed with temporary stop of the acute event; 48 hours after, there was the restart of the bleeding, but was so more severe that caused a consistent anaemization (RBC 2.850.000, Hb 8.1) and that required repeated hemotransfusions and intensive medical therapy. The severity and the lack of autorestriction of hemorrhagic event led us to subject the patient to a surgical operation of Hartmann recto-sigmoid resection with of preternatural anus in left iliac fossa.

Histologic exam of surgical specimen proved massive thrombosis in removed mesenteric vessels.

Postoperative course has been characterized by a quick improvement of hemodynamic conditions, by a gradual overcoming of anaemia and by recovery of performance status. A following echocolor-Doppler showed unmodified the venous thrombosis of the pancreatic and mesenteric vein, with portal compensative cavernomatosis and normodirected flow with high span. Deepened immunologic and hematologic study have excluded other correlations to the development of portal thrombosis.

Discussion

Acute lower gastrointestinal hemorrhage is a relatively uncommon event, valued in approximately 1.5% of all the surgical emergencies; the bleeding is usually self-limiting but, just in almost 10-20% of cases, it's so severe and ingravescient event that requires hospitalization, resuscitation and emergency operation (3). Imdahl, in a literature review, puts on evidence that 80% of acute lower gastrointestinal hemorrhage originate from colon and ano-rectal region. In this last case these are usually hemorrhoids or anal fissures while most frequent colon causes (approximately 60%) are represented by neoplasms, diverticulitis, angiodisplasias, ulcerative colitis with predominant involvement of recto-sigmoid tract (4).

Varices are an uncommon cause of colonic bleeding. Since first report (5), in 1954, until today just 100 cases were reported in literature, and in approximately 75% were referable to portal hypertension due to cirrhosis, biliar atresia, sclerosant colangitis, congestive heart decompensation (6). The cases of primitive colonic varices reported at today are only 22 (25%) (7). These are more usual in male, with mean age of first diagnosis of 41 years (range 14-75). In half cases was involved the total colon, while in the other 50% the elective localization was in the left colon (7). A third of the patients seems to present familiarity, supporting the hypotesis of a genetic factor (autosomic recessive transmission) that determines anomalies of venous plexus (angiodisplastic disorders) (8-11). Lately, in literature, colorectal varices with Klippel-Trenaunay syndrome were described. This is a cutaneous pathology with lone-

some hamartomatous nevi and lower limb varices. We can see shabby forms with absence of the nevi; in these forms lower limb varices are often associated with rectosigmoid varices (12). In none of the case reported in literature there is correlation between idiopathic portal thrombosis that arose up after primitive diagnosis of colon varices, like our case. Idiopathic portal thrombosis, probably in association with coagulative disorders (lack of AT III) or with immunologic disorders (ESL or antiphospholipids Ab syndrome) or with mieloproliferative diseases represents at the same time a common cause of portal hypertension and of variceal bleeding like consequence of porto-systemic shunts (13).

Colonic varices rarely show by bleeding; in a little number of cases repeating of several gastrointestinal hemorrhagic events or the previous positivity in the search of faecal occult blood can be found in anamnesis (6). The trigger of bleeding are unknown; even if some authors give importance to the abrasive action of thick faeces in segment involved from such pathology, this hypothesis seems unacceptable in cases of caecal localization, where faecal material is fluid. The hypothesis of congestion and phlebotasis, to which an alteration of mucosal trophism would be followed with edema, degeneration, ulceration and finally bleeding, seems surely more reasonable (7).

Because of the slight number of reported cases and because of the scattering of many dates it's difficult to draw an univocal diagnostic and therapeutic algorithm; clinical framing and subsequent therapeutic approach rise often up from personal experience rather than well defined guidelines. Before subjecting the patient to complex and invasive exams would have to be excluded hemorrhagic diatesis.

Digestive endoscopy holds an undiscussible role because it's an exam able to identify, also in emergency, cause and site of bleeding and, in expert hands, can guarantee a correct treatment. This procedure is limited by the presence of large coagula (that no can however be removed with relative ease using a colonoscope with large operative channel), from colapsus of varices due to extreme pressure in the course the exam and, finally, from temporary cessation of bleeding, that makes hard to recognize the hemorrhagic source (1).

Dull enema is an unreliable exam because varicose cords can be valued as normal visceral structure or, wrongly, as gas bristles, faecal material, polypoid formations or can be unappreciated at all (14-18).

Scintigraphy could help to distinguish hemorrhagic source (19) such as mesenteric angiography; in particular this last procedure, that it has a very high diagnostic accuracy and a valuable therapeutic potentiality, allows to recognize varices, especially during the

venous phase, with the exception of the cases with excessive dilution of contrast medium just in this phase (20). The main limitation of scintigraphic and angiographic techniques is due to the need of lesion's bleeding to recognize it.

When the diagnosis of colonic varices is formulated, for therapeutic finality too, their characters must be established by giving extreme attention to anamnesis and to possible relationships with pathologies of porto-mesenteric axis, either hypertensive or thrombotic; during this deepening imaging methods must follow a rationale, so the less invasive exam must be executed before complex and unbearable approaches, by starting from US exams (sonotomography and echocolor-Doppler) until angiography when needed.

The treatment is controversial; time by time many therapeutic options are reported either conservative (endoscopic and angiographic) or interventional (colic resection, surgical porto-systemic shunt). Every patient however, based on hemorrhage and performance status severity, must be hospitalized and subjected to intensive medical therapy, by resorting to hemotransfusions and correcting coagulation disorders. While in management of esophageal varices somatostatina-like substances have an important role, in the case of colon varices do not exit trials in such purpose (7).

Between therapeutic options, that can be useful especially in moderate forms, we can signalize endoscopic hemostasis and interventional radiology procedures. Colonoscopy, for the therapeutic options related to the diagnostic ones, can guarantee to control hemorrhage by using several methods, sometimes combined, like electrocoagulation, neodinium YAG laser or argon, the injection of vasoconstrictors or sclerosants substances and, finally by application of ligatures or metal clips (21). The localization of bleeding source is possible by therapeutic maneuvers like embolization or infusion of vasopressine; a permanent catheter can also be used to induce thrombolysis by urokinase in particular when thrombosis of porto-mesenteric axis is associated (22). The rate of colon infarction and perforation after selective embolization is 15%. Another procedure of interventional radiology lately proposed, to treat bleeding of mean entity due to colon varices in portal hypertension, is TIPS (Transjugular Intrahepatic Portosystemic Shunt) (22).

The failure of conservative therapy and the recurrent episodes of bleeding give indication to surgical treatment, that is represented by colonic resection and/or the porto-systemic shunts in the cases of portal hypertension. In our case we made Hartmann colonic resection because of lapsed performance status of the patient (7, 22-24).

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