Review Article

Ectopic varices (a rare cause of stoma bleed)

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Received: 29 August 2014

Accepted: 22 September 2014

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ABSTRACT

Hemorrhage from stoma due to ectopic varices is a rare condition, which can be seen in patients due to portal hypertension of any cause. Resnick et al. were the first to describe in the year 1968. Unfortunately, there has been no randomized controlled trials to formally assess the best management approach for such a condition, and conservative methods are temporary as they re-bleed within 1 year time. Definitive treatment requires a multidisciplinary team approach as they have a high mortality and morbidity rate. The treatment is aimed at reducing the portal-hypertension, which can be achieved with porto-systemic shunts.

Keywords: Stoma hemorrhage, Ectopic varices, Portal hypertension, Porto-systemic shunts

INTRODUCTION

Ectopic varices may be defined as large porto-systemic venous collaterals that can occur anywhere in the abdomen except in the gastro-esophageal region.¹ Though, colostomies, ileostomy and ileal conduits are some of the common sites where ectopic varices are formed and often labeled as stomal varices, other sites such as gallbladder, urinary bladder, uterus, vagina, umbilicus and retro-peritoneum have also been mentioned in the literature by various authors as some of the other rare unusual sites.²⁻⁶

Initially, para-colostomy varices of the stoma was first described by Resnick et al.⁷ in the year 1968, who noted that coincident esophageal varices did not bleed in patients with this type of para-stomal collateral pathway, whereas para-ileostomy varices were first noted by Eade et al.² the year after in patients who had procto-colectomy for inflammatory bowel disease who had developed similar collateral pathways.

PATHOLOGY

It has been hypothesized that stomal varices develop in up to 50% of the patients who have both an external enteric diversion (stoma) and portal hypertension due to any cause, such as in patients with ulcerative colitis with primary sclerosing cholangitis, who had undergone pan-procto-colectomy.^{9,10}

Conte et al.⁸ retrospectively reviewed 72 cases of patients with ectopic varices from the year 1962 to 1989, and concluded that metastatic liver disease was the most frequent cause of para-stoma varices in patients with a colostomy, however the most common predisposing liver condition for portal hypertension was alcoholic liver disease, though as mentioned earlier portal hypertension from any cause can result in para-stomal variceal formation.¹⁰

DIAGNOSIS

In majority of the cases, para-stomal varices can be identified easily on physical examination by a “bluish hue” on the para-stomal skin, which has been named as “caput

medusa,” a term used by both Renick and Eade due to the characteristic appearance of the dilated veins around the stoma.^{8,11}

It is important to note that the only main or sole symptom of this condition is hemorrhage, which is generally caused by local trauma or erosions, which is usually recurrent, painless and profound.^{10,12}

Literature reports that the mean bleeding time from stoma varices after it has been created is between 20 and 36 months; however, overall range of onset can be anywhere between 2 months to 29 years.^{13,14}

In order to confirm the diagnosis of ectopic varices a Doppler ultrasound scan or selective angiography can be performed.¹⁵

MANAGEMENT

Since bleeding ectopic varices from a stoma is a rare condition, there have been no formal randomized controlled trials (RCTs) to assess the best management approach for such conditions,¹⁶ and also, we must not forget that majority of these patients in addition to the above also have esophageal varices; bleeding from which can also divert into the stoma, therefore, making it difficult to identify the primary bleeding point, in fact the source may be in any part of the gastrointestinal tract proximal to the stoma, as a result it has been recommended that in the initial stage, these patients should be managed in a similar manner as we would manage a patient with a gastrointestinal bleed.^{10,14}

Once primary management of bleeding has been achieved, conservative measures include direct pressure (with or without adrenaline soaked gauze), chemical cautery, injection sclero-therapy and suture ligation. It is important to note that there are short-term strategies as variceal bleeds are notorious to re-bleed after sometime.¹⁷

The main advantage of such conservative approach is that it be repeated multiple times with varying amounts of success rates, but they have no role in long-term management as they fail to address the underlying cause of hemorrhage i.e., portal hypertension, therefore, re-bleeding in such cases is common and so is a rule rather than an exception.^{10,16}

Other drastic conservative measure includes muco-cutaneous disconnection and stoma relocation. However, in the review study of Conte et al.¹⁴ this procedure was accompanied by a re-bleeding rate of 100% with sclerotherapy yielding similar results.⁸

In addition to the above, depending on patient factors such as nutrition, disease state and technique used. Each of the above procedure also carries a variable risk of para-stomal ulceration, infection, stricture and necrosis, with general anesthesia and laparotomy worsening the prognosis outcome, leading to prolonged hospital stay.¹⁸⁻²⁰

In an acute bleeding situation where conservative management fail, trans-cutaneous and trans-hepatic embolization has shown to provide adequate control in a high proportion of cases, and hence should be considered first.^{16,21,22} However, they too are temporary measures as it also fail to address the underline cause of bleeding, i.e., portal hypertension; their use is limited by a high rate of re-bleeding at 1 year.¹

Definitive treatment for variceal bleed is aimed at reducing the porto-systemic pressure gradient by a total of 20% or to < 12 mm Hg that can be achieved via porto-systemic shunts. In patients in whom surgery is contraindicated trans-jugular intra-hepatic porto-systemic shunting (TIPSS) is preferred, particularly those in child-pugh class B or C.^{16,23}

Unfortunately, patients with significant heart failure, valvular heart disease and pulmonary hypertension are poor candidates for TIPSS, and in some patients the procedure might not be even possible due to anatomical reasons such as significant portal or hepatic vein thrombosis. In TIPSS, precipitation or worsening of encephalopathy is common and must be taken into consideration when deciding on such procedure.²⁴

It has been recommended that the decision to place a shunt should be made by a gastroenterologist or hepatologist in consultation with an interventional radiologist, and referral to a liver transplant center should be considered in all patients who qualify for TIPSS.^{24,25}

CONCLUSION

Ectopic varices are a rare but clinically significant cause of stoma bleed. They are not only a diagnostic nightmare for the clinicians, but also very difficult to manage due to their rarity, and also there is no RCT data available regarding how best to best manage such patients.

Management includes temporary measures such as: Local pressure (with or without adrenaline), chemical cautery, injection sclerotherapy, suture ligation, mucocutaneous disconnection, and the stoma relocation. Stomal embolization has been found to be useful in controlling bleeding in an acute setting. Definitive treatment involves a multidisciplinary approach where the aim is to reduce portal hypertension, which can be achieved depending on the patient condition and anatomy either by TIPSS or surgical.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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DOI: 10.5455/2349-2902.isj20141103

Cite this article as: Kabir SA, Kabir SI, Pillai S, Chakrabarti M. Ectopic varices (a rare cause of stoma bleed). *Int Surg J* 2014;1:128-30.