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LETTER TO THE EDITOR

Oesophago-gastrectomy in a patient with haemophilia A

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With recent improvements in pre- and postoperative care, major complex surgical procedures are now performed in patients with coagulatory disorders [1]. However, these are seldom reported in the surgical literature. Haemophilia A is an inherited, X-linked, recessive disorder resulting in deficiency of functional plasma coagulation factor (F) VIII and may result in excessive bleeding during and following surgery. The severity of haemophilia, the factor level and the type of surgery will determine the extent of bleeding.

Here, we report a case of a 34-year-old male with haemophilia A who presented with intermittent localized epigastric pain and episodes of belching of 4 months duration. He had no dysphagia, melaena, loss of appetite or loss of weight. The clinical examination was unremarkable.

Upper gastrointestinal endoscopy showed 1 cm polypoid growth at the gastro-oesophageal junction. Multiple biopsies taken under cryoprecipitate cover were reported as adenocarcinoma. A CT scan of the abdomen showed a localized growth in the gastro-oesophageal junction. There was no radiological evidence of metastases.

Since the tumour was small and well localized, the oncological opinion was to go ahead with surgery. Preoperative FVIII assay showed a 30% level, indicating mild deficiency. Preoperative APTT was 42 s (normal 25–40 s) and the Internationally Normalised Ratio (INR) was 1.04. FVIII levels were raised from 30% to 100% by transfusing anti-haemophilic factor (AHF) 30 min prior to surgery. Open Ivor Lewis oesophago-gastrectomy, feeding jejunostomy and a hand sewn anastomosis was performed. The SonoSurg (Olympus Medical Systems Corp., Tokyo, Japan) (ultrasonic dissector/coagulator) was used as the main tool of dissection to minimize bleeding and also to reduce operating time. The total blood loss during surgery was 400 mL. To maintain adequate coagulation during the first 24 h, AHF was transfused, and thereafter, cryoprecipitate was transfused until the fifth post-operative day. Jejunostomy feeding was commenced 6-h postoperatively. The patient was managed in the intensive care unit for 48 h.

Removal of drains, tubes and staples were carried out under AHF and cryoprecipitate cover. Oral feeds were commenced during the second week after a gastrograffin study excluded an anastomotic leak. Histology of the specimen also confirmed the diagnosis of a well differentiated adeno-carcinoma, $pT_1pN_opM_x$.

The patient presented 2 weeks following discharge from the hospital with melaena. Upper GI endoscopy showed oozing from the granulation tissue at the site of anastomosis. Symptoms responded to i.v. omeprazole and administration of AHF. Subsequent endoscopy after 3 weeks showed a well healed anastomosis.

In conclusion, early oesophageal cancer (especially adenocarcinoma) can be cured, if the tumour is excised completely [2]. The same surgical principles should be applied even to patients with major coagulation disorders, provided that pre- and post-operative optimization of coagulation are achieved. During surgery, we used the SonoSurg to minimize bleeding and operating time [3,4]. Bleeding may be further reduced, if minimally invasive surgical techniques are practised [5]. We did not adopt a laparoscopic approach because of lack of expertise in our centre. Therefore, in the present era of modern technology and advanced surgical care, even patients suffering from inherited bleeding disorders, such as haemophilia can be offered major complex surgical procedures with excellent outcome, provided, it is carried out by a team of experienced personnel in centres with adequate facilities.

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