CASE REPORT

MALIGNANT MELANOMA OF THE SMALL BOWEL WITH UNKNOWN PRIMARY: A CASE REPORT

Salman Yousuf Guraya, Muhammad Al Naami, Talal Al Tuwaijri, Maha Muhammad Irfan Arafah*

Division of General Surgery, Department of Surgery, *Department of Pathology, The Division of General Department of Surgery, King Khalid University Hospital, Riyadh, Saudi Arabia

Primary small bowel tumours are rare. However, alongwith stomach it is the most frequent site for metastatic tumours, malignant melanoma being the commonest tumour. The prevalence of malignant melanoma in the gastrointestinal tract without any evidence of a primary lesion in the skin or any other site is extremely rare.

Keywords: Malignant melanoma, Small bowel metastasis, S 100 protein

INTRODUCTION

Although small bowel accounts for 75% of the length and 90 % of the mucosal surface of the gastrointestinal tract (GIT), it is the site of only 3.6% of primary gastrointestinal tumors[1,2]. Metastatic tumors to the upper GIT have an overall prevalence of 1 % to 4 % in postmortem series[3], stomach and the small intestine reported to be the most frequent organs involved[4]. Malignant melanoma is the commonest tumor to metastasize to the GIT, frequent sites of invasion being the small intestine (50 %), colon (31.3 %) and anorectum (25 %)[5]. Between 1 % to 4 % of all the patients with malignant melanoma will have clinically significant GIT involvement diagnosed antemortem, while up to 60 percent of cases with melanoma were found to have metastases at autopsy[6, 7]. This reflects that the majority of such metastatic spread remains asymptomatic thus making the diagnosis a real clinical dilemma.

The incidence of malignant melanoma in the GIT without any evidence of a primary lesion in the skin or any other site is extremely rare[8, 9]. We report a metastatic malignant melanoma in the jejunum with an unknown primary, managed in the surgical unit of King Khalid University Hospital, Riyadh, Saudi Arabia.

CASE REPORT

A 76 year old, previously healthy male, was admitted to our unit through emergency room, seeking medical attention for frequent episodes of fresh rectal bleeding, weight loss, abdominal pain and anorexia of two months duration. His surgical history was remarkable of truncal vagotomy and gastrojejunostomy for peptic ulcer disease about twenty years back. Examination revealed pallor, severe emaciation and tachycardia. Abdominal assessment was essentially unremarkable with normal bowel sounds. Baseline hematological and biochemical profile were within normal range except for gross anemia (Hb: 4.1 gm %) and low albumin (21 gm%). The patient was resuscitated with intravenous fluids and multiple blood transfusions and his nutritional status was improved with total parenteral nutrition for ten days. A high resolution contrast enhanced computed tomography (CT) scan showed about 10x8 cm irregular mass in the proximal bowel with no evidence of pulmonary and pelvic metastases. Small bowel follow through study outlined two well defined filling defects in the proximal jejunum about 15 cm apart showing characteristic "apple-core" appearance (Fig 1). Colonoscopy and upper GI endoscopy could not identify the source of bleeding. Previous gastrojejunostomy facilitated the endoscopist to perform enteroscopy which showed a very friable and necrotic mass in the upper jejunum from which multiple biopsies were taken. The histological report suggested a malignant growth of unknown nature.

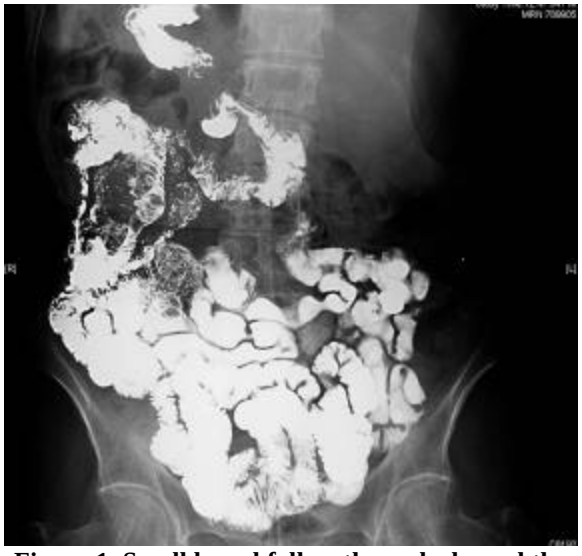

Figure 1. Small bowel follow through showed the characteristic "apple-core" appearance in the proximal jejunum

After optimization of the patient's general condition, he was subjected to exploratory laparotomy through a midline incision. Examination of the abdomen detailed two firm to hard masses in the proximal jejunum about 10 cm apart, proximal 7x7 cm and distal 3x3 cm in dimensions and both projecting into the bowel lumen (Fig 2). There was no evidence of ascites, liver metastasis or lymph node involvement. Resection anastomosis of segment of the jejunum with GIA 50 stapler was undertaken which encompassed healthy bowel margins and wide excision of the mesentry. The final histological report demonstrated metastatic malignant melanoma in both specimens with free resection margins (Fig. 3,4,5). All thirteen lymph nodes were negative for malignancy. An extensive work up including ophthalmologic consultation was sought but could not localize the primary site. A postoperative total body radionuclide scan was unremarkable. Apart from a self limiting transient confusional state, patient made an uneventful recovery and was discharged with a plan for medical oncology review in two weeks.

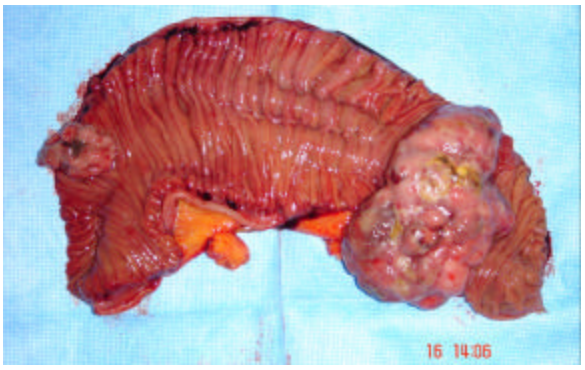

Figure 2. Longitudinal section of the resected specimen of the Jejunum demonstrated metastatic malignant melanomas

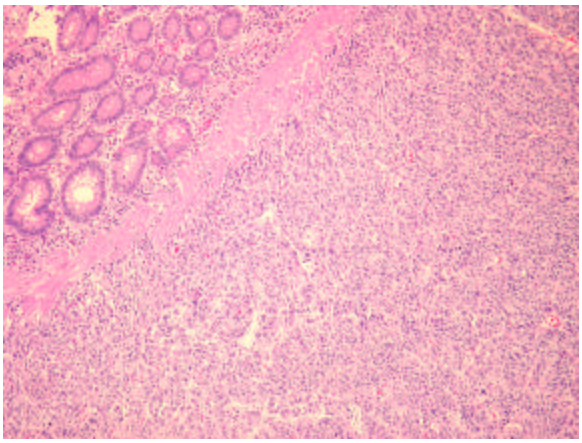

Figure 3. Jejunum, involvement of submucosa by a neoplasm H & E 100X

DISCUSSION

Malignant melanomas account for 1 % to 3 % of all malignant lesions of the GIT[5]. The majority of these cancers represent metastatic spread from a primary site, usually in the skin but may originate from retina, anus, and under the nail[10]. Most patients with GIT metastases by malignant melanoma are asymptomatic and the symptoms, when present, are vague and non specific. Kadakia et al[11] reported anemia in 70 % and acute upper GI bleeding in 50 % of the their patients with metastatic melanomas to the GIT while others documented abdominal pain (60%), bowel obstruction (47%), nausea and vomiting (41%), and GIT bleeding (30%)[12] . Symptoms and signs of acute appendicitis, malabsorption and protein-losing enteropathy may also be the presenting features[13, 14]. An abdominal mass can be identified in only 10% of the patients[15].

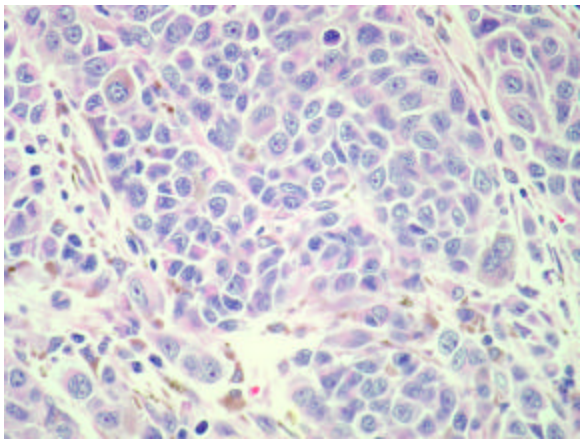

Figure 4. Neoplastic cells with abundant eosinophilic cytoplasm, some of which have brown fine granules H & E 400x

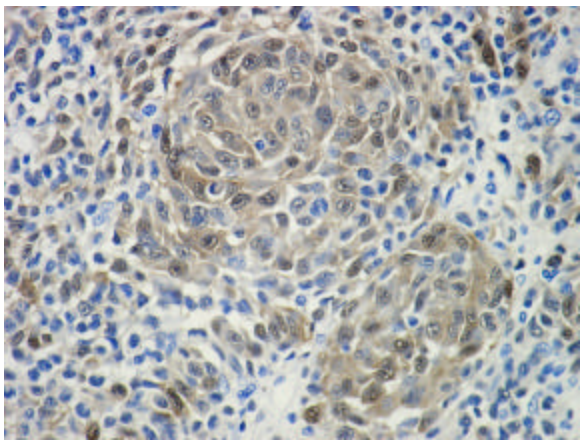

Figure 5. Nuclear and cytoplasmic staining for S 100 protein. Immunohistochemical stain for S 100 protein 400X

Preoperative determination of the extent of small bowel metastases by malignant melanoma is

imperative to improve mean survival time[16]. The relatively low sensitivity of imaging studies (58%) for small bowel follow through and 66% for contrast enhanced CT scans[15] invariably delays the final diagnosis until the resected specimen is examined histologically. Volume-challenged imaging with CT-enteroclysis is the study of choice to detect small bowel melanomas[17]. A high resolution CT scan may reveal mural nodules, "target" or bull's-eye" lesions, excavating masses[18], or diffuse small bowel involvement[5]. Endoscopy may be unrewarding because of the predilection of the metastatic deposits for small bowel serosa and mesentry.

Morphologically, primary malignant melanomas show varying proportions of spindle cells and areas of epitheliod proliferation with abundant large eosinophilic nucleoli and excess cytoplasm[19]. The tumor cells may demonstrate melanin pigment within stromal macrophages but they may be completely amelanotic. Immunohistochemical stains for melanomas that do not depend on melanin pigment include vimentin, S 100 protein and the more specific HMB-45. In contrast, metastatic melanocytes do not exhibit the in situ component but occasionally can exhibit epidermotropism characteristic of a primary tumor.

Curative resection of metastatic malignant melanoma to the GIT remains the mainstay of treatment[18,20], because resection allows a definite diagnosis and, although a cure is rare, can prolong symptom free survival[21]. Surgery should include excision of the tumor with adequate healthy margins along with a subtended wedge of mesentry, to remove the regional lymph nodes[22]. Prognosis is often dismal regardless of the extent of surgery. The average time from the diagnosis of malignant melanoma to the discovery of small bowel metastasis has been reported to be 3.2 years[15]. In our case the primary tumor might have regressed before the appearance of metastasis or was too small to be identified by conventional clinical and laboratory work up.

CONCLUSION

Metastatic malignant melanoma of the small bowel without an identifiable source is exceptionally uncommon. Delayed presentation, non specific symptomatology and low diagnostic yield of the available investigations pose a clinical challenge.

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Address for Correspondence: Dr Salman Yousuf Guraya FRCS, Consultant Surgeon, House No 19 West, Scheme No 2, Farid Town Sahiwal Pakistan. Ph: +92 333 4373635, Fax: +92 42 6279144.
Email: drsyg7@yahoo.com