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Case Report

Small bowel malignant tumor: a diagnostic dilemma, a case report and literature review

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ABSTRACT

A 47 years old Saudi male was admitted to king Khalid hospital with complaints of constipation and abdominal distension. Abdominal examination showed as a case of intestinal obstruction. He was fully evaluated and was treated first conservatively, until all investigative study confirmed as huge small bowel dilatation (mainly jejunum). Exploratory laparotomy confirmed small bowel tumor and on histology an adenocarcinoma. The introduction, history and research paper will also be discussed in this report.

Keywords: Small bowel tumor, Small bowel adenocarcinoma, Jejunal tumor, Jejunal adenocarcinoma, Intestinal obstruction, Small bowel obstruction

INTRODUCTION

A case of small bowel tumor was first described by Hamberger in 1746, as a case with a perforated duodenal carcinoma.[1] The first small-bowel leiomyoma was described by Foerester in 1858 and the first small-bowel leiomyosarcoma was described by Wesener in 1883.[2,3]

An early review of malignant small-bowel tumors was published by Leichtenstern in 1876 and Heurtaux published a review of benign small-bowel tumors in 1899.[4,5]

Small bowel cancer is a rare malignancy that comprises less than 5% of all gastrointestinal malignancies. The estimated annual incidence is 0.3-2.0 cases per 100,000 persons, with a higher prevalence e rates in the black population than the white, and has been recently increasing.[6,7] It is most frequently diagnosed among people aged 55-64, with the incidence increasing after age 40. The current 5-year survival rate in the USA is 65.5%; cancer stage at diagnosis has a strong influence on the length of survival.[8]

Small bowel cancer has four common histological types: adenocarcinoma (30-40%), carcinoid tumor (35-42%), lymphoma (15-20%), and sarcoma (10-15%).[9] Adenocarcinoma of the small bowel (SBA) is most commonly located in the duodenum (57%), while 29% of cases are located in the jejunum and 10% in the ileum.[10] Clinical presentation of small bowel adenocarcinoma (SBA) is nonspecific abdominal discomfort, such as abdominal pain, nausea, vomiting, gastrointestinal bleeding and intestinal obstruction, which leads to an average delay of 6-10 months in diagnosis.[11] Due to the rarity of this cancer, there have been no good screening methods developed for SBA; little is known about the clinical characteristics, treatment modalities or prognosis of patients with SBA, especially in Asians.

The diagnosing of small intestinal malignancy usually delays due to inaccessible of esophagogastroduodenoscopy especially jejunum and ileum causing poor prognostic outcomes. The 40-60% of all patients can be cured and only 32% are non-metastatic lesions.[12] The 5-year survival rates of small intestinal malignant patient is 67.5%.[8]

Standard available diagnostic procedures like abdominal ultrasound, oesophago gastroduodenal endoscopy and colonoscopy enable the diagnosis of only those tumours localized in duodenum, whereas abdominal computed to-138 mography scans (CT scans) do not usually visualize any abdominal mass but only visualize nonspecific changes in the small bowel like swollen lymph nodes or thickening of the bowel wall.

Without any principal diagnosis, with only a suspicion of lesions in the distal segment of small bowel, this then prompts the use of more advanced methods: endoscopy (video capsule endoscopy, double-balloon assisted enteroscopy), imaging techniques using computed tomography (CT) and magnetic resonance; CT/MRI enteroclysis and CT/MRI enterography.

Furthermore, arteriography, scintigraphy and small bowel follow-through are increasingly becoming more important for evaluating small bowel disease.

Despite the wide spectrum these aforementioned diagnostic methods we are still missing a specific screening method that could simultaneously meet the following criteria: cost, availability, precision of intestinal lumen evaluation together being able to take samples for pathological examination, and invasiveness. Despite these aforementioned procedures being all performed in Poland, they are unfortunately only readily available usually at highly specialized medical centers.

CASE REPORT

A 47 years old Saudi male presented without personal or family history of digestive diseases, cancer or genetic disorders presented to the emergency department. He complained of increasing diffuse abdominal pain over 12 h with nausea, vomiting and constipation.

The patient denied any history of weight loss. He was afebrile, hemodynamically stable with moderate diffuse abdominal tenderness and distension.

Six months ago, he experienced an episodic attack of distending pain in his left lower quadrant, nausea and vomiting; he was treated conservatively in a local hospital. However, his symptoms were not completely relieved, and were later aggravated.

Abdomen was distended with normal bowel sounds suggestive of incomplete obstruction. Clinically no mass was palpated. He was given a trial for conservative management abdominal ultrasonography initially revealed hugely dilated small bowel loops.

Plain X-ray abdomen showed dilated loops of small intestine without any fluid levels (figure1).

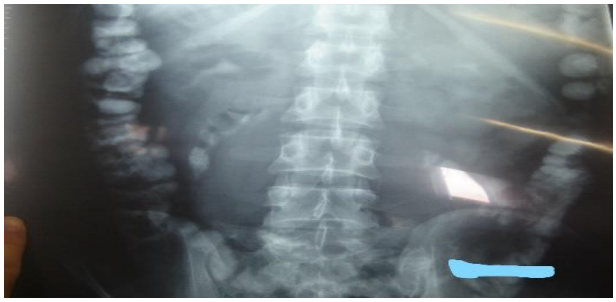

Figure 1: Small bowel dilatation.

CT scan reported as homogenous soft tissue mass with filling defect and contrast enhancement in small bowel mostly in jejunum (figure 2 and 3).

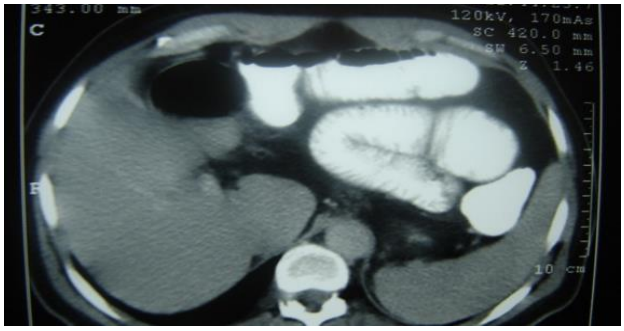

Figure 2: Small bowel dilatation.

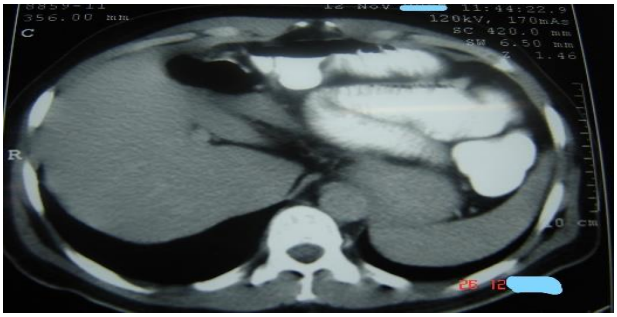

Figure 3: Another view of small bowel dilatation.

Due to patient's state deteriorating and aggravated abdominal pain, a decision for emergency surgery was made.

The patient underwent exploratory laparotomy, intra-op findings revealed-the small intestine was dilated with a thickened and overworked wall extending almost the whole length up to the obstacle. A tumor was found 10 cm from duodenojejunal junction partially obstructing the lumen with multiple small tumor nodules in almost whole of jejunum, mesentery and anterior abdominal wall. Omentum was studded with multiple small nodules. Liver and pancreas were free. There was no ascites (Figure 4A, 4B, 6A, 6B).

Resection anastomosis of jejunum was done and around 60 cm of jejunum was resected (figure 5) with side to side anastomosis.

The specimen was sent for HPE which reported as adenocarcinoma of small bowel with tumor infiltrating the whole wall with evident serosal nodule and infiltration to anterior abdominal wall. Margins were free and 1 out of 2 lymph nodes was found positive with capsular invasion.

He had a past history of road traffic accident in 1982 with fracture in right femur post plate fixation and left kidney stone removal in 1972. No past history of similar complaints. No other family history and co morbidity.

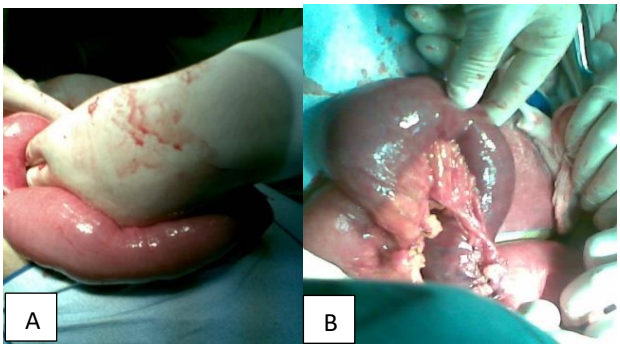

Figure 4: (A) Dilated small bowel. (B) Another view of the dilated small bowel.

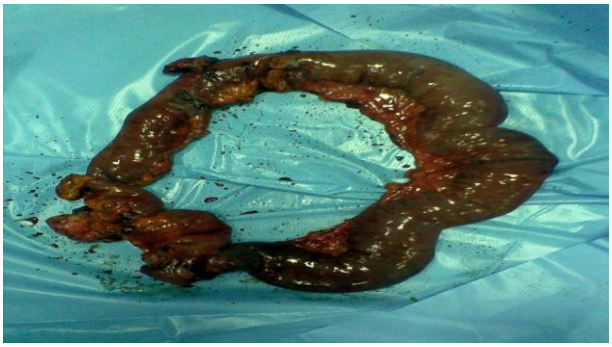

Figure 5: Resected Specimen.

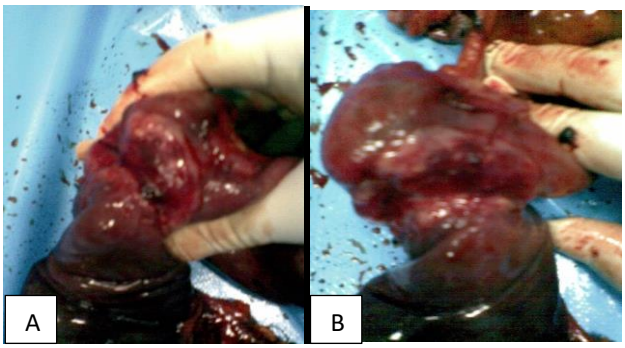

Figure 6: (A) Jejunal tumor mass. (B) Another view of mass in jejunum.

DISCUSSION

Whereas the small bowel represents 75% of the length of the digestive tract and 90% of the absorptive mucosal surface area, tumor of the small bowel is rarer than other gastrointestinal malignancies. The possible explanations include the high levels of IgA and the more rapid transit in the small bowel compared to the large bowel. Little bacteria and more sensitivity to stress in the small bowel also contribute to the low tumor incidence.[13] Though small bowel cancer normally occurs in elderly patients, in this case, it was found in a 26-year-old young man. The mass remained undetectable until he had an incomplete small bowel obstruction with lymph node metastasis. This was similar with studies, in which diagnosis of SBA was mainly obtained at advanced stages; ~40% of patients have lymph node metastasis (stage III), and 35 to 40% have distant metastasis (stage IV).[11,14]

The symptoms of SBA are initially nonspecific abdominal discomfort; diagnosis is delayed and usually in the context of emergency involving an occlusion (40%) or bleeding (24%), which is similar to the presentation of our patient. For diagnosis of SBA, CT scans have an overall accuracy rate of 47%.[11,15] While CT scans can detect the lesions, they cannot provide precise data of the intestinal mucosa and miss some small or flat lesions. The PET/CT technique is being used to differentiate small intestinal malignant tumours from benign ones. The uptake of F-FDG is related to tumor size, infiltration and lymph node metastasis; the higher the uptake of F-FDG, the higher the tumor invasiveness.[16,18] Gastroscopy and enteroscopy can be appropriate if the tumor is located close to the proximal duodenum or far from the terminal ileum. The rest of the small bowel cannot be accessed without the use of video capsule endoscopy (CE) or DBE (double balloon enteroscopy). The definite diagnostic yield of CE is only 20-30%, while DBE accounts for 60-70% of the diagnostic yield for intestinal diseases.[17] However, CE is suitable for diagnosing scattered, small and multiple lesions, as well as active bleeding; it is convenient, non-invasive, secure and comfortable. In contrast, the DBE procedure is uncomfortable, less tolerated and difficult to complete; these factors influence its diagnosis.[17] A baseline plasmatic CEA and CA 19-9 assay is necessary, especially in cases of advanced disease because CEA and CA 19-9 levels are of prognostic value.[18]

Surgical resection with clear margins and regional lymph node resection remains the treatment of choice in localized SBA; indeed, it is often required even in metastatic SBA due to the high probability of obstruction or severe hemorrhage.[19] To date, there has been no standard chemotherapy regimen against SBA. Several studies have explored the role of palliative chemotherapy in advanced SBA. Hong et al have shown in stage IV patients who received palliative chemotherapy that overall survival (OS) increased significantly compared to those who did not receive chemotherapy (8 vs.

3 months, p=0.025).[12] Ecker et al have shown that median OS was superior for patients with resected stage III SBA who were receiving chemotherapy versus those who were not (42.4 vs 26.1 months, p<0.001). As for the Asian population, Mizyshima et al showed that, in patients with non-curative resection or unrespectable distant metastasis, the response rate to chemotherapy was 31.6%, and the 3-year OS rate was significantly higher compared to the response rate without chemotherapy (26.3 vs. 13.8%; p=0.008).[20,21] Several chemotherapy drugs have also been evaluated in the treatment of metastatic SBA. Zaanan et al have shown that the median OS in advanced SBA patients treated with FOLFOX was 17.8 months, the longest survival among different chemotherapy regimens.[18] Two phase II studies have been conducted to evaluate the efficacy of different chemotherapy regimens in advanced SBA: the response rates were around 50%, the median progression-free survivals 7.8 and 11.3 months and the median OS 15.2 and 20.4 months.[22,23]

Newer agents, such as endothelial growth receptor (EGFR) antibody drugs, and newer combinations are being explored as the second line for improved treatment of advanced SBA.[24] From limited clinical reports, a combination of fluoropyrimidine with platinum compounds (FOLFOX or CAPOX) has been proposed as the first-line treatment for palliative chemotherapy in metastatic SBA treatment.[25] In view of the results of genetic studies, the patient underwent palliative chemotherapy for eight cycles of FOLFOX and was doing well as of his last follow-up.

CONCLUSION

The presentation of tumor of the mid GIT is often very nonspecific, making diagnosis a real difficulty. Only small bowel enema in experience hands can give diagnostic information. The surgeons and clinicians must have a high index of suspicious in their mind of this problem diagnosis in nonspecific non diagnostic abdominal symptoms to improve the diagnostic accuracy and overall survival of patient. The clinicians must be willing for extra diagnostic work including even an exploratory laparotomy to diagnose small bowel tumor at an early stage. Surgical resection is the only curative option for small intestinal adenocarcinoma.

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