CASE REPORT

Adenocarcinoma of DJ flexure presenting as Wilkie's Syndrome – A rare case report.

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Abstract :

Adenocarcinoma of duodeno jejunal flexure is a rare entity, the presenting features of which are vague and non specific. We report a case of a 51 years lady, with a presentation of pain abdomen, vomiting and weight loss. A diagnosis of Wilkie's syndrome was made on CECT Abdomen, based on a narrow aorto mesenteric angle, but laparotomy revealed a stricturous lesion at the duodeno jejunal flexure. A segmental resection of the lesion and anastomosis was performed, and the histopathology confirmed the diagnosis as adeno carcinoma of the duodeno jejunal flexure region. It was further concluded that the Wilkie's syndrome had resulted from the reduced mesenteric fat pad caused by the duodeno jejunal flexure carcinoma and cancer cachexia.

Keywords : Adenocarcinoma of duodeno jejunal flexure, Ligament of Treitz, Segmental resection, Wilkie's syndrome

Introduction:

Adenocarcinoma of the duodeno jejunal flexure is an extremely rare condition and its presentation is delayed due to vague symptomatology and difficulty in establishing the diagnosis. It poses an unique challenge in terms of pre operative diagnosis, treatment plan and post operative management.

It is usually diagnosed by CT Abdomen and intra operative findings. Due to the vague symptomatology the condition is usually detected in a locally advanced stage or with distant metastasis. Segmental resection of the duodeno jejunal flexure is the surgical treatment of choice. Post operative Chemotherapy improves the performance status.

Wilkie's syndrome also known as superior mesentery artery syndrome, cast syndrome, chronic duodenal ileus, vascular compression of the duodenum and aorto mesenteric duodenal compression, is a rare condition of high intestinal obstruction due to the narrow angle between the abdominal aorta and the superior mesentery artery causing compression of the 3rd part of the duodenum.

Here in is a report of a rare case of adenocarcinoma of duodeno jejunal flexure presenting as Wilkie's syndrome.

Case Report:

A 51 years lady presented with complaints of postprandial epigastric pain, vomiting, anorexia and loss of > 10% of weight in 3 months (rapid weight loss). Physical examination revealed- poor nourishment, mild abdominal distension and decreased bowel sounds. Her weight was 40 kg and height was 1.54m (body mass index = 16.8 kg/m). Laboratory investigations revealed- anemia, hypoproteinemia and electrolyte imbalance, which were corrected pre operatively. Plain X-ray abdomen showed a grossly distended stomach with Ryles tube, reaching the pelvis [Fig-1].

Abdominal ultrasonogram revealed grossly distended stomach. Upper GI endoscopy revealed-Lax LES, dilated body of the stomach, dilated D1 & D2.Contrast CT scan revealed- distended stomach and distended 1st and 2nd parts of duodenum with compression of the third part of the duodenum [Fig-2,3].

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How to cite this article:
Rajababu Pakanati , Ramarao K. V. & Lalityaswarna Pethakamsetty : Adenocarcinoma of DJ flexure presenting as Wilkie's Syndrome – A rare Case Report. International Journal of current Medical and Applied sciences; 2016, 12(2),96-98 .

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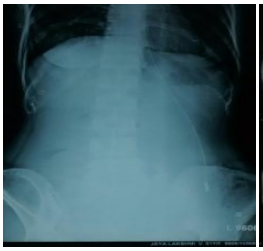
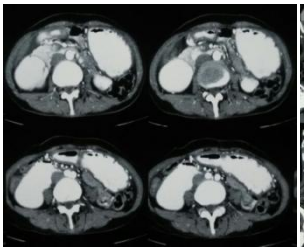
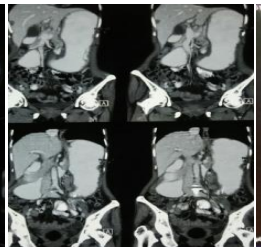
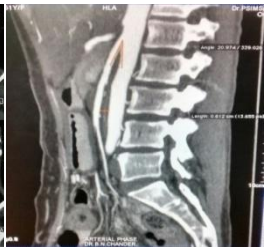

Figure 1: *Figure 2:* *Figure 3:* *Figure 4:*

Figure 1 : Plain X-Ray Abdomen showing grossly distended stomach with ryle's tube, reaching the pelvis.
Figure 2 : Contrast CT Scan showing distended stomach and distended 1st and 2nd parts of duodenum.
Figure 3: Coronal view of contrast CT Scan showing distended stomach and distended 1st & 2nd parts duodenum.
Figure 4: Contrast CT scan showing decreased angle between the Abdominal aorta and the Superior mesenteric artery.

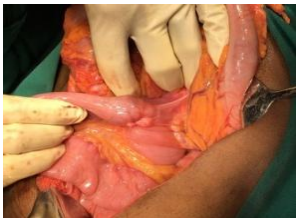
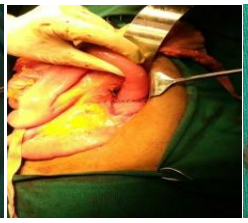
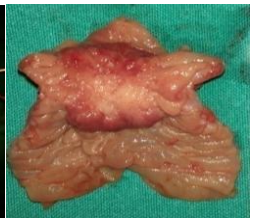
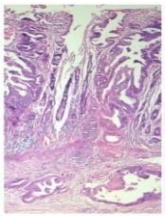
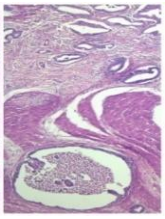

Figure 5 : *Figure 6:* *Figure 7:* *Figure 8A* *Figure 8B*

Figure 5 : Stricture at DJ flexure at the level of Ligament of Trietz
Figure 6 : Duodeno jejunal anastomosis following segmental resection of the stricturous lesion.
Figure 7: Cut surface of the specimen showing grey white circumferential ulceroinfiltrative growth.
Figure 8: (A) Duodenal mucosa with tumour glands infiltrating the lamina propria (LP). (B) Duodenal muscularis propria (MP) infiltrated by tumour glands (H and E, x 100)

Aortomesenteric distance of < 7mm (Normal: 10 to 28 mm) and aortomesenteric angle of < 21° (Normal:25° to 60°) was noted [Fig-4]. With a diagnosis of Wilkie's syndrome, a laparotomy was done which revealed a stricturous lesion in DJ flexure, at the ligament of trietz[Fig-5]. Segmental resection of the duodeno jejunal flexure including the stricturous lesion was done with a margin of 1.5cm on duodenal side and 5 cm margin on jejunal side. Duodenojejunal anastomosis(end to end), and a distal feeding jejunostomy was done [Fig-6].

Macroscopic examination revealed a grey white circumferential ulceroinfiltrative mass, in the resected specimen, measuring 3.5 cm (inner circumference) x 1 cm (length) x 0.5 cm (depth), at the stricture area causing subtotal (moderate) luminal compromise [Figure 7].

Microscopic examination of the sections from duodenal mass revealed transmurally infiltrating well differentiated adenocarcinoma [Figure 8]. TNM: T4,N0,M0, Grade I, Stage IIB.

The postoperative course was uneventful. Subsequently she was started on chemotherapy. Patient was doing well on follow up after one month and a weight gain of 5 kgs was noted.

Discussion:

Malignant tumours of small intestine are very rare accounting for 0.1 to 0.3% of all malignancies and 1% of all gastrointestinal malignancies [1].

Adenocarcinomas account for 36.9% of all small bowel malignancy with the remaining comprising of carcinoid tumours (37.45%), lymphoma (17.3%) and stromal tumours (8.4%) [2].

Duodenal adenocarcinoma , accounts for < 0.4% of all GIT tumours and 56% of all duodenal malignancies [2,3]. True incidence of adenocarcinomas occurring at ligament of treitz is unknown. Due to their vague and nonspecific presenting symptoms, a delayed diagnosis or misdiagnosis is common. Adenocarcinomas usually present in the 6th and 7th decades. The most common presenting complaint is intermittent pain due to partial intestinal obstruction. Other presentations included anorexia, weight loss, abdominal distension, jaundice and diarrhoea [3,5]. The mean duration of symptoms before presentation is 10 months (range, 0-24 months) [4].

In our case the patient presented with symptoms of subacute small bowel obstruction, the cause of which was initially discerned as Wilkie's syndrome based on contrast enhanced computed tomography (CT).

Wilkie's syndrome is an uncommon syndrome, with an incidence of 0.1 to 0.3%, characterised by compression of the 3rd part of duodenum between the superior mesenteric artery and aorta resulting in recurrent mechanical duodenal obstruction [8]. Any condition that narrows the aortomesenteric angle can precipitate entrapment of 3rd part of duodenum resulting in Wilkie' syndrome, which is also known as superior mesenteric aretery syndrome, cast syndrome, chronic duodenal ileus, vascular compression of the duodenum and aorto mesenteric duodenal compression.

The condition most commonly affects underweight individuals with a history of rapid weight loss as in - catabolic states such as cancer, surgery, trauma, burns, or psychiatric problems, dietary disorders like anorexia nervosa (or) malabsorption. Confirmation of

Wilkie's syndrome requires radiographic procedures such as upper gastrointestinal series, hypotonic duodenography and contrast CT scanning. The CT criteria for the diagnosis of WS are aortomesenteric angle of < 22° and aortomesentric distance of < 8mm which were evident in the current case [9,10]. More recently EUS and PET scan are also options for diagnosis. Successful treatment of WS requires identification and removal or reversal of the precipitating cause. Initial conservative management, comprising of adequate nutrition and nasogastric decompression, is recommended. Following failure of the above treatment, surgical procedures like duodenojejunostomy, gastrojejunostomy or division of Ligament of Treitz may be required.

In the present case, in addition to causing subtotal obstruction, the carcinoma of the duodeno jejunal flexure region resulted in cachexia with consequent reduction in mesenteric fat pad culminating in Wilkie's syndrome, which further exaggerated the symptoms of upper GIT obstruction.

Duodenal first and second part tumors are treated by Whipples' procedure. For resectable cancers of third and fourth part of duodenum, segmental resection is the treatment of choice [11]. Duodenojejunal segmentectomy is the treatment of choice for duodeno jejunal flexure tumors with lymph node clearance, though lymph node positivity does not preclude resection, and the prognosis of these tumors is good. Survival by stage is 65 % for stage I, 48 % for stage II, 35 % for stage III, and 4 % for stage IV [12].

In patients with R0 resection, adjuvant chemotherapy will result in an improvement in the disease-free survival, and overall survival in stage I, II, and III in small bowel adenocarcinomas. Post-op 5 -FU is the mainstay chemotherapy. FOLFOX (oxaliplatin, 5-FU, and leucovorin) and FOLFIRI (irinotecan, 5-FU, and leucovorin) regimens significantly improve the performance status and progression-free survival in the treatment of metastatic small bowel adenocarcinoma [13,14].

Conclusion:

Primary duodeno jejunal adenocarcinoma, although very rare should be considered as the differential diagnosis of small bowel obstruction, when evaluation of the upper and lower GI tract is unremarkable or misleading. Computed tomography scan has a high accuracy in detecting the disease and its metastatic spread, to stage the disease. In this case, Duodeno jejunal adenocarcinoma is considered as the etiological factor of Wilkie's syndrome due to obstruction close to DJ flexure causing tumor induced cachexia. Resection and anastomosis of DJ flexure lesion was performed.

Segmental resection is the treatment of choice in duodeno jejunal adenocarcinoma and the aim of surgery is to achieve tumour free margins and limit post-op morbidity. For unresectable tumours gastro jejunostomy is the treatment of choice.

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Conflict of interest: None declared
No source of funding.