Malignant Tumor at D-4 Mimicking Wilkie's Syndrome

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Abstract Primary carcinoma of the duodenum is rare. Here we present a case of megaduodenum due to duodenal adenocarcinoma mimicking Wilkie's syndrome which was managed by resection and anastomosis.

Keywords Duodenal adenocarcinoma · Megaduodenum · Wilkie's syndrome

Introduction

Wilkie's syndrome is an atypical case of high intestinal obstruction often posing a diagnostic dilemma. Though rare, it is a well recognized clinical entity affects young adults. The syndrome is also known as Superior Mesenteric Artery Syndrome, Cast Syndrome, Chronic Duodenal Ileus and Aorto-mesenteric Duodenal Compression. It is commonly caused by compression of 3rd part of duodenum at the narrow angle between superior mesenteric artery and aorta. Compression of 3rd part of duodenum by peritoneal bands or shortened ligament of Trietz may also be rarely responsible. Malignant tumor of distal duodenum is not included in the etiology of Wilkie's syndrome but can present as high intestinal obstruction.

Herein is a report of a rare case of malignant tumor of distal duodenum mimicking Wilkie's syndrome

Case History

A 60 year old gentleman presented with the history of recurrent bilious vomiting, weight loss, postprandial fullness

for the last 2 months. Examination revealed anemia and a hugely dilated stomach. Endoscopy showed dilated stomach and duodenum. Barium series showed megaduodenum with abrupt cut off at the 3rd part of duodenum (Fig. 1). CT scan confirmed obstruction at the 3rd part of duodenum with no mass lesion suggesting a diagnosis of superior mesenteric artery syndrome. The patient was prepared by gastric lavage, correction of anemia, malnutrition, electrolyte imbalance. Surgery was planned in the form of diagnostic laparoscopy and bypass if SMA syndrome was confirmed.

Laparoscopy detected a stenosing lesion in the 4th part of duodenum at the angle of Trietz (Fig. 2). The lesion was assessed and mobilized laparoscopically proximally till the SMA was seen. After full mobilization we suspect it might be a malignant tumor. Subsequently open exploration was done and the tumor resected out along with a segment of intestine with macroscopic clear margins which was confirmed by frozen section. The tumorous lesion was also malignant on frozen section. The portion of intestine resected included 5 cm of duodenum proximal to tumor and 10 cm of jejunum distal to it with mesentery and adjacent nodes. G.I. continuity was restored by hand sewn end to end duodeno-jejunostomy in two layers. A distal feeding jejunostomy was done. Postoperative recovery was uneventful. The histopathology suggested a diagnosis of Grade II adenocarcinoma with infiltration upto serosal fat. Resection margins were free. There was no lymph node metastasis noted among all six lymph nodes. There was no perineural or angiolymphatic invasion. At one year follow-up the patient is doing well.

Discussion

Though SMA syndrome was first described way back in 1861 by Rokitsky, it was in 1921 that Wilkie provided a

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Fig. 1 Barium series shows megaduodenum with abrupt cut-off at 3rd part

patho-physiological description of the condition and named it “Chronic Duodenal Ileus” [1]. Wilkie later published one of the largest series, totaling 75 cases. The incidence of this condition varies from 0.013–0.3% of the barium series of upper GI tract [2]. But many have suggested that the condition is actually over diagnosed because of its confusion with other causes of megaduodenum and one of the cause of megaduodenum is malignant stenosing lesion of distal duodenum, usually adenocarcinoma.

The most common cause of Wilkie’s syndrome is thought to be a sharp decrease of aorto-mesenteric angle from the normal 45 degree to the range of around 6–16 degree resulting in the extrinsic compression on duodenum as it passes between aorta and SMA [2]. Patients usually present with chronic vague abdominal symptoms, recurrent episodes of pain, bloating, bilious vomiting, weight loss and anemia. Typically symptoms are relived by prone or knee-elbow position. The condition is difficult to diagnose and made by process of exclusion. Barium series of upper GI tract, CECT abdomen and Upper GI Endoscopy can help in diagnosis. Surgical treatment in the form of duodenojejunostomy to bypass the obstructed portion gives excellent result. The same is not true for the cases when the cause of megaduodenum is malignant lesion at distal duodenum and usually requires surgical resection as in our case.

Duodenal adenocarcinoma is rare, accounting for only 0.35% of all gastrointestinal carcinomas, and carcinoma of the fourth part of the duodenum constitutes approximately 10% of all duodenal adenocarcinoma [3]. Tumor of distal duodenum usually presents in similar pattern as of Wilkie’s

syndrome but patients are usually in the sixties. Barium series of upper GI tract, CECT scan, Upper GI Endoscopy and intraoperative finding helps in making the diagnosis. The treatment is surgical resection, which in the 2nd part of duodenum may require pancreaticoduodenectomy. A review of the literature indicates that survival, which is related to nodal status, the grade of the tumor, and surgical results, seems to be longer in patients with carcinoma in this part of the duodenum compared to those with carcinomas in other parts of the duodenum. Moreover, segmentary resection has been reported to achieve a favorable outcome [3].

Primary duodenal carcinoma is a distinct entity with a better prognosis than pancreatic cancer after radical resection. It favors the descending duodenum and is closely linked with villous adenoma and epithelial dysplasia [4].

Radical surgical resection of these tumors, even by segmental resection, provides a more favorable prognosis for duodenal carcinoma than for pancreatic tumors [5]. The method of treatment is pancreaticoduodenectomy for the upper part and segmental resection for the distal part of duodenum [6]. Adjuvant therapy after resection for duodenal cancer has not been well studied however many patients were treated with 5-FU-based therapy. Nevertheless there is no clear cut evidence that chemotherapy prolongs survival [7]. Sixty-six patients with primary adenocarcinomas of the duodenum recorded by the Swedish Cancer Register during 1958–1973 are reviewed. The mean age was 66 years, and female:male ratio 1.2:1.0. The predominant symptom was duodenal obstruction. Correct diagnosis was made in 68% by conventional barium meal examination. Hypotonic duodenography and duodenoscopy are necessary diagnostic aids. In 25% of the patients the diagnosis was first made at postmortem examination. Thirty-two patients had metasta-

Fig. 2 Laparoscopic view of growth at D-4 before mobilization

ses at first diagnosis. Forty-three per cent were radically operated and 43% palliatively. The overall one-year survival was 67% and five-year 18%. There was a tendency for longer survival time for patients with more distally situated carcinomas. Duodenopancreatectomy gave a longer survival time than duodenal resection [8].

We have done segmental resection for our case and at the end of one year follow-up he is doing well and this is also supported by Lowell et al from there study that survival of patients with adenocarcinoma of the distal duodenum is surprisingly good, and segmental resection is the procedure of choice [9].

Conclusion

Wilkie's syndrome is a rare cause of upper gastrointestinal obstruction in adult and should be considered when other causes have been excluded. Surgical intervention in the form of surgical bypass (duodenojejunostomy either open or laparoscopic method), in the event of failure of conservative treatment, gives gratifying results. Malignant obstruction of distal duodenum should be kept in mind while dealing with any elderly patient presenting like

Wilkie's syndrome and surgical resection of the involved segment gives good long term result.

References

1. Wilkie D (1921) Chronic duodenal ileus. *Br J Surg* 204–214
2. Ylinen P, Kinnunen J, Hockerstedt K (1989) Superior mesenteric artery syndrome: a follow-up study of 16 operated patients. *J Clin Gastroenterol* 11:386–391
3. Yildirim S, Culhaoglu AB, Ozdemir N (1995) Carcinoma of the fourth part of the duodenum: report of a case. *Surg Today* 25 (12):1034–1037
4. Scott-Coombes DM, Williamson RC (1994) Surgical treatment of primary duodenal carcinoma: a personal series. *Br J Surg* 81 (10):1472–1474
5. Wellner I, Banga P, Haulik L (2001) Surgical resection of tumors in the distal duodenum. *Magy Seb* 54(4):215–218
6. Bakaeen FG, Murr MM (2000) What prognostic factors are important in duodenal adenocarcinoma. *Arch Surg* 135(6):635–642
7. Dabaja BS, Siki D, Pro B (2004) Adenocarcinoma of small bowel: presentation, prognostic factors, and outcome of 217 patients. *Cancer* 101:518–526
8. Alwmark A, Andersson A, Lasso A (1980) Primary carcinoma of the duodenum. *Ann Surg* 191(1):13–18
9. Lowell JA, Rossi RL, Munson JL, Braasch JW (1992) Primary adenocarcinoma of third and fourth portions of duodenum. Favorable prognosis after resection. *Arch Surg* 127(5):557–560