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Wilkie Syndrome: Rare Cause of Obstruction (A Case Report and Literature Review)

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Authors' contributions

This work was carried out in collaboration among all authors. Author AEB designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors MB and EAL managed the analyses of the study. Author EAL managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Acute mesenteric clamp syndrome or Wilkie's syndrome is one of the rare causes of bowel obstruction. It is the result of the compression of the third duodenum by a vascular clamp formed by the superior mesenteric artery and the aorta after disappearance of the perivascular fatty tissue. The functional signs are non-specific and are common to other diseases of theupper digestive tract and are dominated by recurrent post prandial abdominal pain, nausea and vomiting and weightloss.Physical examination is poor. The management of The aorto-mesenteric clip syndrome includes medical treatment and surgical treatment. The aim of this work is to high light the diagnostic difficulties of high intestinal occlusions in general and of the aorto-mesenteric clip syndromein particular, its clinic, possible complications and management.

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Keywords: Proximal bowel obstruction; superior mesenteric artery; Willkie's syndrome; gastro-jejunostomy anastomosis.

1. INTRODUCTION

The aorto-mesenteric clamp syndrome or Wilkie's syndrome is one of the rare causes of small bowel obstruction (high intestinal occlusion). Gastrointestinal tract disordersdue to this anomaly occurin 0.013 to 0.3% of the general population [1]. It is the result of compression of the third duodenum by a vascular clamp formed by the up permesenteric artery and the aorta after disappearance of perivascular fatty tissue [2]. The clinical signs are mainly dominated by recurrent postprandial abdominal pain, nausea and vomiting and weightloss, abdominal distension with dyspeptic syndrome, epigastric tenderness [3]. We present the case of a 32-year-old patient admitted to our visceral surgical emergency department for high haemorrhagic obstruction following aorto-mesenteric forceps.

2. CASE PRESENTATION

We're talking about a 32-year-old patient, chronic smoking, who's been vomiting for two months, regurgitation complicated three days ago by an occlusive syndrome made up of material and gas shutdown, without externalized digestive bleeding the whole evolving in a context of apyrexia and alteration of general condition. General examination found a ptient conscious, with normocoloured conjunctiva.

Examination of the abdomen notes a general abdominal sensitivity, fasting and a distension, of the abdomen without palpable mass, no hepatomegaly or splenomegaly. Hernial orificesareas and lymphnode arenormal.

3. PARA CLINICAL ASSESSMENT

Unprepared Abdomen

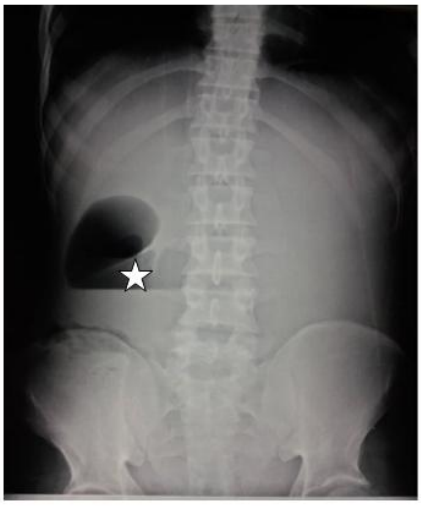

Fig. 1. Dilatation of the duodenum (asterisk) and presence of a hydroaerous level opposite the right flank of the 12th dorsal vertebra

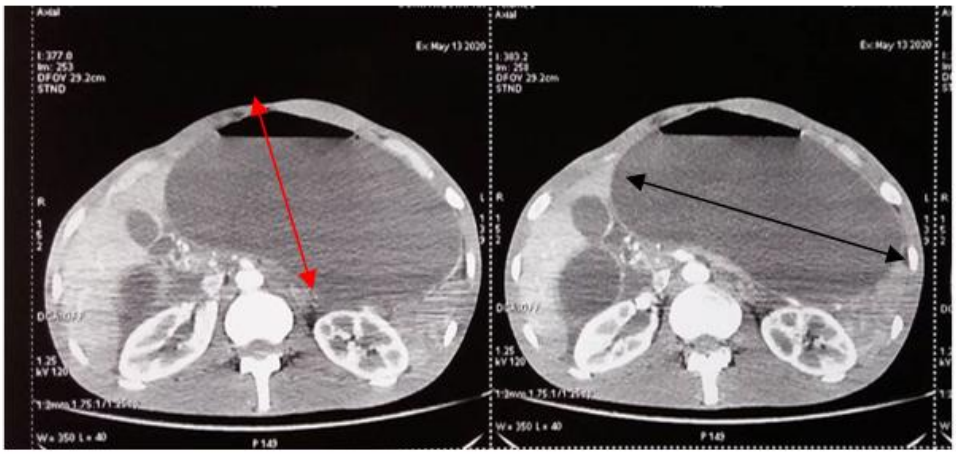

Fig. 2. Axial view of the CT abdomen in arterial phase shows dilatation of the stomach and the first and second part of the duodenum with significant narrowing of the third part of the duodenum between the superior mesenteric artery and the aorta

Thoraco-abdominal CT scan: Significant gastric distension occupying the entire abdominal cavity. This distension is responsible for the compression of adjacent structures. Appearance in favor of aorto-mesenteric clamp syndrome.

Complete blood count : hemoglobinat =17.2 g/dl, WBC =21180/mm, platelet =286000/mm, Na+ = 134 mEq/l, K+ = 4.8mEq/l, Urée = 0.74 g/l, Créat= 18.2 mg/L, CRP= 24 mg/L, ALB =56 g/L

The patient benefited froma gastro-jejunal bypass on a handle with a sub-mesocolic Omega Atypical gastric resection of a pre-perforative necrosis area of 8 cm long axis on the anterior surface of the stomach Left sub-phrenic drainage/SS, Drainage of Douglas/DR CDS

The patient was placed supine, under general anaesthesia, intubated and ventilated, nasogastric tube in place, a urinary catheter placed, under 0.75 mg norepinephrine. The incision wasmedial, straddling the umbilicus.

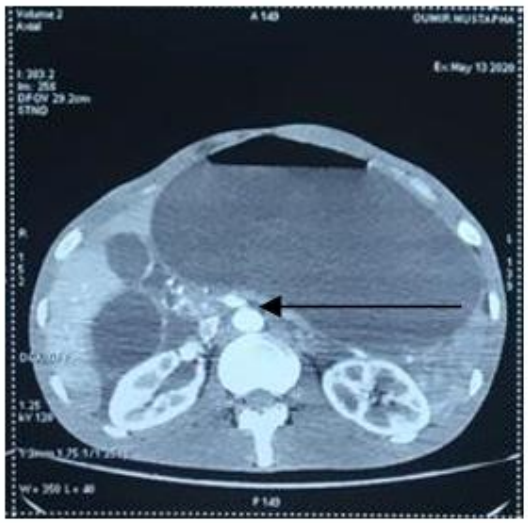

Fig. 3. Compression of the 3rd duodenumwith dilatation of the rest of the duodenal framework and the stomach (arrowhead)

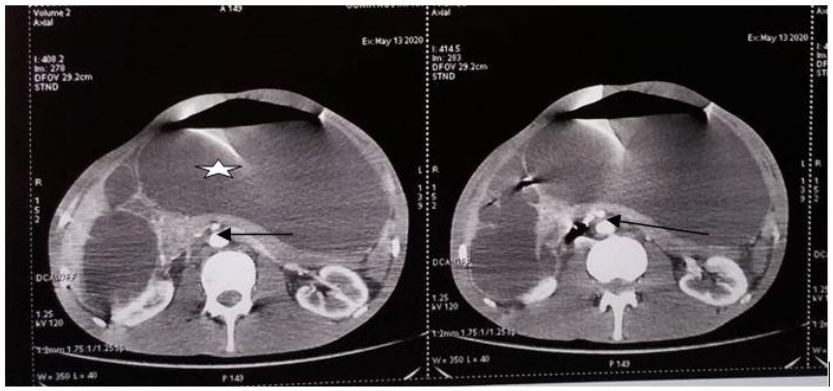

Fig. 4. Dilation of the stomach with displacement of structures; note the compression of the right kidney by the dilated 2nd duodenum (asterisk) and the aorto meenteric forceps (arrowhead)

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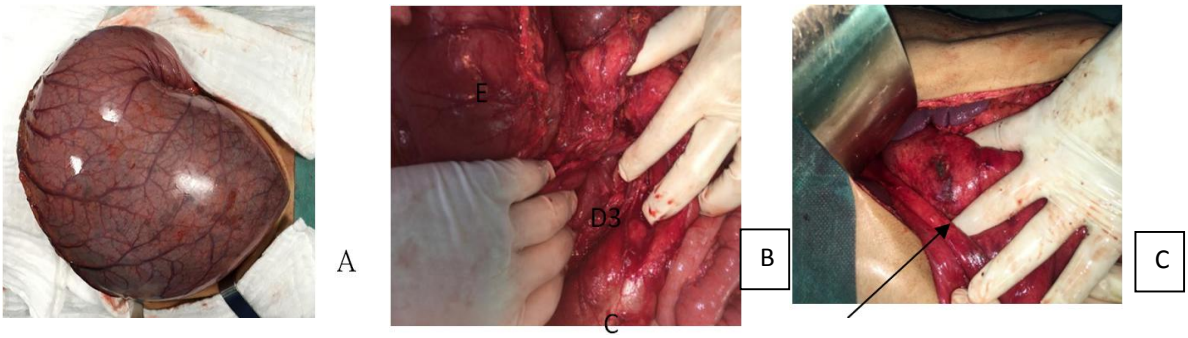

Fig. 5. Intraoperative images : A: gastric distension; B: Duodenumpinchedbetween the aortaposteriorly and the SMA (E=stomach; D3=3rd duodenum; C=colon;) C: preperforative areas of the stomach

Exploration found a significantgastric distension, DI, DII, DIII upstream of the mesenteric vessels, producing an aorto-mesenteric forceps on the DIII with a zone of constriction. The rest of the duodenal frame work, hail and collapsed colon Area of localized necrosis on the anterior surface of the stomach body

Atypical gastric resection of the anterior surface of the stomach body with an 8 cm long axis carrying thepre-perforativelesions

4. DISCUSSION

Aorto-mesenteric clamp syndrome is one of the rare causes of bowel obstruction. It was first described by Rokitanskyin 1842 and the first case series of this disease was published by Wilkiein 1927, whichled to the disease also being called Wilkie syndrome [4]. Aorto-mesentric clamp syndrome is more common in women, children and adolescents [5].

Pathophysiology can be attributed to all factors that contribute to a reduction in the angle between the aorta and the superior mesenteric artery (SMA) (by 6-16°) and a distance (by 2-8 mm) resulting in a high obstruction [6]. The most known cause being congenital anomalies, the aorto-mesenteric clamp syndrome has also been linked to other causes including: rapid weight loss following bariatric surgery, scoliosis or after abdominal surgery including total coloprotectomy with ileoanal Reservoir [7]. AORTO-MESENTERIC CLAMP SYNDROME is also linked to severe and disabling diseases such as cancers, malabsorption syndrome, AIDS, severe trauma and burn victims [4]. For our patient, there is no obvious etiology that would have caused the AORTO-MESENTERIC CLAMP SYNDROME according to the investigations made and the assessments. Since the patient came to us in a state of altered general condition with no notion of previous spinal or colorectal surgery and no notion of recent trauma.

Functional signs are not specific and are common to other diseases of the digestive tract. Post prandial abdominal pain, abdominal bloating, early satiety, bilious nausea and vomiting are observed. These signs mayvary depending on whether the duodenal obstruction is partial or complete and maybelimited to simple postprandial abdominal pain or maymanifest as early postprandial bilious food vomiting accompanied by weight lossif the obstruction is severe [8]. Physical examination is poor, even for thin patients, exceptin case of abdominal distension or intestinal occlusion [6,9]. Our patient experienced the same symptoms for 2 months, resulting in weight loss and abdominal distension following a stasis stomach. Confirmation of duodenal compression and dilatation and gastric stasis is crucial to differentiate Aorto-mesentric clamp syndrome fromo ther disorders such as gastro paresis, functional dyspepsia, cyclic vomiting syndrome and eating disorders and this is done through imaging [7]. The barium transit shows the dilatation of the duodenum and its pinching [10]. To establish the diagnosis, most authors consider the maximum threshold of 25° (normal values 38-56) at this angle and a maximum distance of 8 mm (normal values 10-22) between the superior mesenteric artery and the aorta on CT scan.

Management of THE AORTO-MESENTERIC CLIP SYNDROME includes medical treatment and surgical treatment consisting of a gastro jejunal bypass to circumvent the obstruction. The goal of the medical treatment is to regain weight and at the same time regain the fat around the root of the superior mesenteric artery. Enteral nutrition can beused by a jejunostomy tube made beyond the treitz ligament, or by a nasogastric tube [9].

The medical treatment of AORTO-MESENTERIC CLAMP SYNDROME consists of the insertion of a nasogastric tube and aspiration of the stomach and duodenum, placing the patient in a left-lateral position, and above all compensating for hydro electrolytic disorders, and instituting a hypercaloric double diet, enteral by a naso-jejunal and parenteral tube. Success in this case is around 72% but with recurrences of around 30%.

Failure of medical treatment is pronounced when there is no improvement in symptoms. There is no time limit for failure, however, treatments hould be continued for between 2 and 12 days, although one successful treatment lasting 169 days has beenreported in a child [2]. To date, there is no scientific evidence that sets a time limit for medical treatment, and the duration of medical treatment varies from 9 to 45 days depending on the paediatric series [11]. To date, there has been no scientific data that suggests the appropriate time limit for the duration of medical management or the optimal period before surgery [1,6]. The median durations of medical treatment have been shown to be 9 days

(range, 2-62 days) and 65 days (range, 13-169 days) in a pediatric case series [5,10]. In our study, the median duration of treatment was 45 days

The surgery consists of a derivation by gastro jejunostomy or duodeno-jejunostomy, which can be performed laparoscopically, or modify the anatomical conditions by mobilizing and decreasing the duodeno-jejunal angle by positioning the jejunum to the right of the SMA after sectioning the Treitz muscle according to Strong's procedure, the best results obtained are those of the duodeno-jejunal anastomosis. Gastro-jejunal anastomosis is effective on gastric distension, but to a lesser degree on the duodenum, giving a disappearance of vomiting but persistence of epigastric fullness, as compared to Strong's procedure, it is not feasible in all patients because of adhesions and duodenal distension with recourse in case of a second failure to a gastro-jejunal anastomosis or a latero-lateral duodeno-jejunal anastomosis [7,2]. SMAS is a rare condition, which is why there are no randomized clinical trials comparing different surgical techniques. Duodenal jejunal anastomos is was introduced by Starley n 1910 and has been the most widely used technique with a success rate of 90%, how ever this technique is not applicable to all patients [12]. A few isolated cases and series in which patients were treated laparoscopically with SMAS have been reported, demonstrating good results with this method and the advantages of laparoscopy [6]. For our patient, the medical treatment was the medical treatment was unsuccessful. So we traied a surgical treatment by a gastro-jejunal bypass.

5. CONCLUSION

Aorto-mesenteric clamp syndrome is rare and can occuratanyage. Diagnosisis made by a combination of clinical and radiologicalsigns. Treatment begins with medical measures including decompression of the stomach with nasogastric tube, feeding and correction of hydroelectrolytic deficits, if thereis no response, surgery should be considered. In the event of any high obstruction in a severely undernourished patient, the diagnosis of SMAS should be made.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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