Coexistent Superior Mesenteric Artery Syndrome and Nutcracker Syndrome: A Case Report and Review of Literature

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Abstract: Background: Superior mesenteric artery syndrome or Wilkie’s syndrome is a rare condition consisting of external compression of the third part of duodenum between the Superior mesenteric artery and the aortorachidian plan resulting in duodenal obstruction. The vascular compression of the duodenum is a very rare condition exceptionally associated to Nutcracker syndrome: an uncommon vascular abnormality which is caused by external compression of the left renal vein. Case presentation: We report a case of a 27-year-old man admitted to our department with a history of severe weight loss, post prandial epigastric abdominal pain with anorexia and occasional vomiting over the past two years. He reported two episodes of macroscopic hematuria. Physical examination revealed a cachectic patient with a body mass index of 14kg/m². The abdomen was tender with distension. Laboratory studies revealed a mild anemia and hypoalbuminemia with no other abnormalities. An abdominal CT scan showed a massive gastric and dilated proximal duodenum, with the compression of the third portion of the duodenum between the aorta and the Superior mesenteric artery. In addition, the left renal vein was compressed by the Superior mesenteric artery. The diagnosis of Superior mesenteric artery syndrome associated to Nutcracker syndrome was made. Conservative treatment (correction of electrolyte imbalance, enteral nutrition) fails. The patient was treated surgically with gastrojejunostomy in order to gain weight and to reduce pain. Conclusions: Superior mesenteric artery syndrome and Nutcracker syndrome are uncommon vascular abnormalities, exceptionally associated. The diagnosis is most of the time a challenging and must be considered in the differential diagnosis of gastro-intestinal obstruction and unknown cause of hematuria. CT findings could be helpful for the diagnosis. Most of the time the conservative treatment with adequate nutrition will result in weight gain. Surgical option can be considered when conservative treatment fails.

1. Introduction
Superior mesenteric artery (SMA) syndrome or Wilkie’s syndrome is a rare condition consisting of external compression of the third part of duodenum between the SMA and the aortorachidian plan resulting in duodenal obstruction. Nutcracker syndrome (NCS) is an uncommon vascular abnormality which is caused by external compression of the left renal vein (LRV) either between the aorta and the SMA or the aorta and the spine. We report through this case a rare coexistence of SMAS and NCS with atypical vascular compression of the left renal vein. The angle between the abdominal aorta and the SMA or the aorta and the spine can be considered when conservative treatment fails.

2. Case Description
A 27-year-old man was transferred to our department for management of significant weight loss of about 20 Kg associated to abdominal pain. Two years ago, he complained of post prandial abdominal pain with anorexia and occasional vomiting. He reported two episodes of macroscopic hematuria. Physical examination revealed a cachectic patient with a body mass index of 14kg/m². The abdomen was tender with distension. Blood tests revealed a mild anemia and hypoalbuminemia with no other abnormalities. An exhaustive etiological inquiry was thus initiated. The biological screening of an infectious diseases (tuberculosis, whipple diseases, hepatitis B and C, human immunodeficiency virus infection and digestive parasitosis), an inflammatory pathology (celiac disease, systemic lupus erythematosus, systemic scleroderma, vasculitis, anti phospholipids syndrome), a hormonal dysfunction (dysthyroidism, adrenocortical insufficiency) and tumor markers, were all negative. Endoscopic gastroduodenoscopy was normal. An abdominal CT scan showed a massive gastric and dilated proximal duodenum, with the compression of the third portion of the duodenum between the aorta and the Superior mesenteric artery (Fig 1 A). In addition, the left renal vein was compressed by the Superior mesenteric artery. The angle between the abdominal aorta and SMA was 16,4°(Fig 1B,C). The diagnosis of Superior mesenteric artery syndrome associated to Nutcracker syndrome was made. The patient received initially a conservative treatment (correction of electrolyte imbalance, enteral and parenteral nutrition with prophylactic low weight molecular heparin). In view of the persistence of abdominal pain, our patient was transferred to our department with a history of severe weight loss, post prandial epigastric abdominal pain with anorexia and occasional vomiting. He reported two episodes of macroscopic hematuria. Physical examination revealed a cachectic patient with a body mass index of 14kg/m². The abdomen was tender with distension. Blood tests revealed a mild anemia and hypoalbuminemia with no other abnormalities. An exhaustive etiological inquiry was thus initiated. The biological screening of an infectious diseases (tuberculosis, whipple diseases, hepatitis B and C, human immunodeficiency virus infection and digestive parasitosis), an inflammatory pathology (celiac disease, systemic lupus erythematosus, systemic scleroderma, vasculitis, anti phospholipids syndrome), a hormonal dysfunction (dysthyroidism, adrenocortical insufficiency) and tumor markers, were all negative. Endoscopic gastroduodenoscopy was normal. An abdominal CT scan showed a massive gastric and dilated proximal duodenum, with the compression of the third portion of the duodenum between the aorta and the Superior mesenteric artery (Fig 1 A). In addition, the left renal vein was compressed by the Superior mesenteric artery. The angle between the abdominal aorta and SMA was 16,4°(Fig 1B,C). The diagnosis of Superior mesenteric artery syndrome associated to Nutcracker syndrome was made. The patient received initially a conservative treatment (correction of electrolyte imbalance, enteral and parenteral nutrition with prophylactic low weight molecular heparin). In view of the persistence of abdominal pain, our patient was transferred surgically with gastrojejunostomy in order to gain weight and to reduce pain. No treatment was necessary for NCS, in view of normal renal function and the patient was discharged with outpatient follow-up.
3. Discussion

The SMA syndrome is a rare anatomic variant, consisting in compression of the third portion of duodenum between aorta and SMA. The incidence is estimated at 0.1% to 0.3%. SMA syndrome preferentially occurs in females than males with a general age range of 30 to 40 years old but it can occur at any age1,2.

Clinical presentation may be acute, realizing high gastrointestinal tract obstruction, or chronic, with intermittent post prandial abdominal pain, nausea, vomiting with abdominal distension, weight loss and early satiety1,2. Diagnosis is based on abdominal CT scan, showing the gastric and proximal duodenal dilatation with compression of third portion of duodenum and especially a shortened aortomesenteric distance. It allows for measurement of aortomesenteric angle. Recent reports advocated that the angle<8mm and a narrow aortomesenteric angle<22° are suggestive of the diagnosis 7.

Initial treatment is usually conservative based on the correction of electrolyte imbalance, enteral or parenteral nutrition. Hyperalimentation increases the mesenteric fat pad, thus increasing the aortomesenteric angle and improving symptoms. Surgical intervention may be considered if conservative treatment fails. Several techniques are possiblesuch as the release of the ligament of Treitz, duodeno-jugal or gastro-jugal anastomosis, repositioning of jujenum to the right of SMA. Laparoscopic duodenjejunostomy is preferred due to reported success rates of more than 80%, it solves, at the same time, gastric and duodenal distention 1,3.

Our patient presents another exceptional compression syndrome of the left renal vein. The NCS is caused by the compression of LRV between aorta and SMA, resulting in renal venous hypertension. Its prevalence is still underestimated. Clinically, patients can be asymptomatic or present left flank pain (43-65%), microscopic or macroscopic hematuria (8-69%), orthostatic proteinuria (4-26%), varicoceles (8-21%), severe pelvic congestion leading to infertility or surrenal insufficiency signs4,5. Positive diagnosis is based firstly in doppler ultrasound, which is a helpful and noninvasive diagnostic tool. A ratio of peak systolic velocity of the aortomesenteric segment to the hilar portion of>4.2 to 5is considered one of the diagnostic criteria of NCS. CT scan and Magnetic resonance imaging (MRI) are also required to confirm the diagnosis of NCS, SMA branching angle of <35 degrees from the aortic origin can be useful for the diagnosis of NS. On the other hand, a pressure gradient of >1 mm Hg during venography has been found to be significant and helpful in confirming this diagnosis6.

He management of NCS depends in the severity of the clinical presentation. A conservative approach with surveillance may be sufficient for old patients or those with mild symptoms. Surgical options should be considered in symptomatic patients. Several surgical options have been described. These include direct renocaval reimplantation, resection of fibrous tissue and placement of a synthetic wedge at the aortomesenteric angle, LRV transposition SMA transposition, nephropey, renal autotransplantation, gonadocaval bypass, laparoscopic splenorenal venous bypass, and laparoscopic left gonadal vein ligation8,9.

LRV transposition seems to be the most commonly performed surgical intervention for the NCS, with a good long-term results. Another management option is percutaneous endovascular stenting of the LRV, which have been used with success as well as long-term resolution of symptoms. However, LRV stent migration, restenosis or thrombosis have been rarely reported7. SMA and nutcracker syndromes are two rare vascular compression conditions. Concurrent occurrences is exceptional. We searched the literature for previous case reports regardingMEDLINE. We performed also, additional reviews based on the literature citations of the identified articles. Only 11 cases were reported in the literature6,9-10. The summarized cases are shown in Table 1.

There were more male than female (9/3), the mean age was 29.75 years, ranged from 15 to 62 years, The mean BMI was 17 kg/ m2 (range 11.7 to 22 kg/m2). Commonest presentation was abdominal pain and vomiting, only one Tunisian Patient presented with hematuria and the second one is our patient. Hematuria was reported as the most common symptom of NCS, it is due to elevated LRV pressure resulting in the rupture of varices secondary to venous congestion and hypertension in the collecting system. It is reported in only Tunisian patients, this is may be related to ethnic anatomical variation. Three patients required operation : gastro jejunostomy for our patient and the patient reported by OH MJ and the third one was from Tunisia and reported by Jomni et al. The rest improved on conservative treatment.

In conclusion, we reported a case of coexistence of SMA and NC Syndromes. Concurrent occurrences is exceptional, only 11 cases reported in the literature. The management of NCS depends upon the clinical presentation and the severity. We should consider conservative treatment, because of the high rate of spontaneous resolution.

References

Conservative treatment with NA: Not Available, BMI: Body Mass Index

Table 1: Literature Reporting of Superior Mesenteric Artery Syndrome combined with Nutcracker syndrome

<table>
<thead>
<tr>
<th>Reference</th>
<th>Sex/age</th>
<th>BMI (Kg/m²)</th>
<th>Symptoms</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boursou et al ³</td>
<td>F/29</td>
<td>11.7</td>
<td>Abdominal pain/No US</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Vulliami et al ⁴</td>
<td>M/55</td>
<td>17</td>
<td>Epigastric pain, vomiting/bleeding/ No US</td>
<td>NA</td>
</tr>
<tr>
<td>Oh MJ ⁵</td>
<td>M/23</td>
<td>21</td>
<td>Abdominal pain, vomiting/bleeding/No US</td>
<td>Duodeno-jejunostomy</td>
</tr>
<tr>
<td>Nunn et al ⁶</td>
<td>F/19</td>
<td>NA</td>
<td>Epigastric pain, vomiting/No US</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Jonni et al ⁷</td>
<td>F/35</td>
<td>22</td>
<td>Anemia, gastrointestinal obstruction/ Hematuria</td>
<td>LRV transposition</td>
</tr>
<tr>
<td>Inal et al ⁸</td>
<td>M/23</td>
<td>NA</td>
<td>Epigastric pain, vomiting and bleeding/No US</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Michael et al ⁹</td>
<td>M/28</td>
<td>17</td>
<td>Epigastric pain, vomiting and bleeding/No US</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Pivawer et al ¹⁰</td>
<td>M/15</td>
<td>NA</td>
<td>Abdominal pain, vomiting/No US</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Alenezy et al ¹¹</td>
<td>M/17</td>
<td>16.9</td>
<td>Abdominal pain, vomiting/No US</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Iqbal et al ¹²</td>
<td>M/62</td>
<td>NA</td>
<td>No specific symptoms</td>
<td>Conservative*</td>
</tr>
<tr>
<td>Kim et al ¹³</td>
<td>M/24</td>
<td>17</td>
<td>Vomiting, left flank pain</td>
<td>Conservative*</td>
</tr>
<tr>
<td>This case</td>
<td>M/27</td>
<td>14</td>
<td>Abdominal pain, vomiting/Hematuria</td>
<td>Gastrojejunostomy</td>
</tr>
</tbody>
</table>

NA: Not Available, BMI: Body Mass Index

*Conservative treatment with nutritional support (parenteral/enteral Nutrition)

US: Urinary symptoms, LRV: Left renal Vein

Figure 1: (A) Sagittal CT scan showing the reduction of the angle (16.4°) between aorta et superior mesenteric artery, (B) Axial CT scan showing duodenal compression (D3) between the abdominal aorta and superior mesenteric artery, (C) Axial CT scan showing dilated left renal vein owing to superior mesenteric artery compressing.