The superior mesenteric artery (SMA) originates from the anterior aspect of the abdominal aorta, just below the celiac trunk at the level of the L1 vertebra. Normally, the SMA is surrounded by fat and lymphatic tissues which provide a cushion between it and its surrounding structures. The mesenteric fat pad between the SMA and aorta forms a natural angle between them, called the aortomesenteric angle. The normal aortomesenteric angle is 25-60°. The aortomesenteric distance, the distance between aorta and SMA, is usually 10-28mm. Loss of this cushion of fat can lead to acute angulation of the SMA and reduction in the aortomesenteric distance. This in turn can cause extrinsic vascular compression of other structures by the SMA, giving rise to two distinct vascular compression disorders: 1) **SMA syndrome (Wilkie’s syndrome)** – compression of the third part of the duodenum by the SMA. 2) **Renal vein entrapment (Nutcracker syndrome)** – compression of the left renal vein by the SMA. SMA vascular compression disorders are thus triggered by any condition that diminishes the normal fat cushions, causing angulation and reduction in distance between the aorta and SMA. Fat cushion loss is commonly seen under conditions of severe weight loss – anorexia nervosa, hypercatabolic states (burns, trauma, malignancy, major surgery) and malabsorption. Various surgical procedures, eg bariatric surgery, scoliosis surgery, ileoanal pouch anastomosis and intestinal resection can cause extrinsic vascular compression of other structures by the SMA, giving rise to two distinct vascular compression disorders: 1) **SMA syndrome (Wilkie’s syndrome)** – compression of the third part of the duodenum by the SMA. 2) **Renal vein entrapment (Nutcracker syndrome)** – compression of the left renal vein by the SMA.

**Superior mesenteric artery syndrome**

SMA syndrome (also known as Wilkie’s syndrome, aortomesenteric compression, arteriomesenteric duodenal compression or duodenal vascular compression) was originally described by Von Rokitansky in 1842, but reported in the literature by Wilke in 1927. 

- **Clinical presentation**
  
  Symptoms associated with SMA syndrome are vague and non-specific. They commonly include abdominal distension and bloating, epigastric pain, nausea and vomiting. The pain is characteristically relieved on lying prone in a left decubitus position. This is explained by the fact that the mesentery and small bowel move from the right side of the abdomen to the left, widening up the aortomesenteric angle. Patients may present acutely with signs and symptoms of duodenal obstruction or they may complain of chronic abdominal pain which may last for months or even years.

**Radiographic findings**

- **Plain radiograph:** Demonstrates dilated, fluid and gas filled stomach.

- **Barium studies:** Shows dilated first and second parts of the duodenum with an abrupt transition point in the third part (figure 1). Other signs include extrinsic compression of the third part of the duodenum and collapsed small bowel distal to the compression.

- **Computed tomography (CT):** Contrast enhanced CT is the imaging modality of choice today. It can be used to reach the correct diagnosis immediately. An abdominal CT does not only allow direct visualisation of the obstructed bowel in relation to the SMA, but can also demonstrate narrowing of the aortomesenteric angle and reduction of the aortomesenteric distance. In addition, other causes of obstruction can be excluded (figure 2).

**Treatment**

Treatment is initially conservative with a high success rate. Conservative measures commonly include gastric decompression via a nasogastric tube, fluid-electrolyte balance and nutritional support, which is provided either with total parenteral nutrition or feeding via a nasojejunal tube placed in the proximal jejunum. Manoeuvring patients to prone left-sided decubitus position during meals may also be helpful. Oral feeding is encouraged after the initial weeks and once the patient gains weight. Surgery is considered for symptomatic patients if conservative treatment fails or if weight gain does not improve symptoms. There is no consensus regarding the duration of conservative management and this is left to the discretion of the treating clinicians. Surgical options include Treitz ligation with mobilisation of the duodenum, gastrojejunostomy, duodenal derotation, duodenojejunostomy and SMA transposition surgery. Duodenojejunostomy is the most common procedure and with advancements in laparoscopic techniques, laparoscopic duodenojejunostomy offers a minimally invasive therapeutic option. It is also effective in majority of the patients.

**Left renal vein entrapment**

Left renal vein entrapment refers to the extrinsic compression of the left renal vein (LRV) between the SMA and aorta, resulting in obstruction and dilatation of the vein. In the literature the condition is commonly called the Nutcracker syndrome (NCS) or Nutcracker phenomenon (NCP), and these terms are used interchangeably. However, NCP refers to the anatomic and haemodynamic findings whereas NCS is reserved for patients manifesting clinical symptoms of the abnormality. LRV compression was first reported by El-Sadr and Mina in 1950. The term NCS was coined by the Belgian physician De Schepper (1972).

**Clinical presentation**

Several clinical features are associated with NCS. Most common is haematuria which can vary from micro to macrohaematuria. This is caused by rupture of the thin walled
veins into the collecting systems due to renal vein hypertension. Haematuria may or may not be associated with flank pain.

The next common symptom is pain. As a result of the LRV compression and venous reflux, prominent collateral veins may develop, causing pelvic congestion (figure 3). This in turn can present as gonadal vein pain syndrome, characterised by abdominal or flank pain with dyspareunia, dysmenorrhoea and varicocele. Varicoceles almost always occur on the left side.

Mild to moderate proteinuria is also commonly seen.

Radiographic findings

Intravenous urogram: Often normal, but in some cases notching of the renal pelvis and ureters due to extrinsic compression from collateral vessels may be seen.

Doppler ultrasound (DUS): Real-time DUS is recommended as the first diagnostic test for LRV entrapment, even though agreed diagnostic criteria are not established. Measurements of the LRV AP diameter and peak velocities using DUS aids in the diagnosis of LRV entrapment. Average normal LRV diameter is 4-5mm with a transition in calibre at the level of extrinsic compression. In LRV entrapment, stenosis of the LRV occurs at the aortomesenteric portion with distension of the vein. This increases the ratio of the renal hilum diameter to the aortomesenteric diameter. DUS is also used to measure the pressure gradient. Normally the pressure gradient between LRV and inferior vena cava is less than 1mmHg. A gradient of 1mmHg or more indicates LRV hypertension.

Computed tomogram (CT): CT demonstrates narrowing of the aortomesenteric angle and LRV compression between the aorta and SMA (figure 4). Associated collaterals can also be easily visualised.

Treatment

Treatment options depend on the severity of symptoms and age of the patient. Conservative approach with observation and routine urinalysis for at least two years is recommended in patients with mild haematuria or those under 18 years of age. It is thought that with formation of collaterals, LRV hypertension improves and symptoms may resolve. Use of ACE inhibitors is also advocated by some in reducing proteinuria associated with LRV entrapment.

Surgery is only carried out when strongly advocated. Surgery may be considered when symptoms are severe or persistent, and if they fail to resolve after 24 months of conservative treatment. Surgical options include LRV transposition, SMA transposition, renal vein-to-IVC shunt, varical ligation, gonadal vein bypass and even nephrectomy for persistent haematuria. Recently, endovascular surgery using endovascular stent graft placement has been applied.

Conclusion

SMA compression disorders are uncommon but well documented worldwide. These conditions may be encountered by clinicians in a variety of specialities and therefore, despite the low incidence, clinicians should be aware of these compression disorders. With an increasing number of people with anorexia nervosa and bariatric surgery these disorders may manifest themselves as atypical complications with increasing frequency. The symptoms encountered in SMA compression disorders are non-specific and overlap with other clinical conditions, making recognition and diagnosis difficult. As a result, patients suffer for a considerable amount of time before the condition is diagnosed and treatment started. In patients with sudden or severe weight loss presenting with common symptoms of compression (post-prandial epigastric pain, bloating, nausea and vomiting in SMA syndrome; haematuria, pelvic pain and varicocele in LRV entrapment) there should be a high suspicion of SMA compression disorders.

Acknowledgments

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References

Figure 1A
Water soluble contrast meal shows a normal calibre stomach, first and second parts of duodenum, however contrast fails to enter the third part.

Figure 1B
This is the same patient’s CT abdomen examination carried out two weeks prior to the water soluble contrast meal. It shows a markedly distended and fluid filled stomach and proximal duodenum. The transition point is within the third part of the duodenum.

Figure 2
Water soluble contrast study in a 69-year-old who presented with weight loss and persistent vomiting shows pooling of the contrast within the antrum and duodenum, at the junction of second and third parts. This raised a suspicion of SMA syndrome, but a subsequent abdominal CT showed circumferential constriction with wall thickening at the second part of duodenum in keeping with a stenosing tumour (white arrow).
Figure 3
Multiple varicosities in the left pelvis (white arrow) and left flank (black arrow) in a patient with LRV entrapment.

Figure 4A
Compression of the LRV between the abdominal aorta and SMA in a 42-year-old who presented with weight loss. Note the dilated LRV.

Figure 4B
Sagittal view showing narrowing of the aortomesenteric angle.