Entrapment Syndromes

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Abstract
Entrapment syndromes represent a pathological process that vascular specialists encounter infrequently. However symptomatic patients are often young with impaired quality of life and successful treatment can produce great benefit, making knowledge of these conditions essential.

The purpose of this review was to bring together the entrapment syndromes to understand and gain consensus on the aetiology, pathogenesis, diagnosis and modern management of these rare and interesting vascular disorders. This includes entrapment syndromes of the popliteal artery, superior mesenteric artery, coeliac artery, renal vein and iliac vein.

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Introduction

Most modern vascular practice encompasses atherosclerotic arterial disease and venous insufficiency. External compression of normal vessels represents a different and infrequent pathology. The best known is thoracic outlet syndrome, which was recently reviewed.1 This review focuses on the remaining compression syndromes as shown in Fig. 1. Compression syndromes can cause ischaemia (arterial or visceral entrapment) or stasis (venous entrapment). Many patients are asymptomatic but symptomatic patients are often young with impaired quality of life and successful treatment can produce great benefits.

Methods

The Pubmed and Medline databases were searched from 1966 to 2007 to identify articles relating to vascular entrapment syndromes. The terms 'popliteal entrapment', 'superior mesenteric artery syndrome', coeliac artery entrapment', 'iliac vein entrapment', 'May Thurner’s', 'nutcracker' and 'renal vein entrapment' were used.

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Results

Popliteal artery entrapment syndrome

Popliteal artery entrapment syndrome (PAES) results from an abnormal anatomical relationship between the popliteal artery and its surrounding musculotendinous structures. First described in 1879, the first operative treatment was reported 80 years later (medial head of gastrocnemius myotomy and popliteal artery endarterectomy). 

Epidemiology
PAES predominantly affects young men (male:female ratio 9:1) lacking atherosclerotic risk factors. The true incidence is unknown. 3.8% of post-mortem limbs have aberrant anatomy consistent with PAES. A prevalence of 0.165% in Greek soldiers suggests that clinically significant PAES is less common than the anatomical abnormality. PAES is responsible in 60% of young patients with ischaemic pain. Unilateral PAES should prompt contralateral investigation as bilateral PAES is reported in 22-67%.

Pathophysiology
Recurrent popliteal artery compression causes intimal damage, thrombosis, distal embolisation, post-stenotic dilation and true aneurysm formation, seen in 13.5% of all cases. Unlike atherosclerosis, entrapment features neovascularisation, inflammatory cell infiltrate and vessel wall disruption, stimulating fibrosis and collagenisation. Once aneurysm or thrombus formation occurs, arterial damage is irreversible.

Embryology and classification
The types of popliteal artery entrapment are differentiated on an anatomical basis (Table 1) and are better understood in the context of embryological development. The proximal popliteal artery results from the fusion of the ischiadic artery and the developing femoral arterial plexus, whilst the midportion is derived from the axial artery. The distal third of the popliteal artery is only formed from the fusion of the anterior and posterior tibial arteries once the medial head of gastrocnemius migrates to occupy a medial position in the popliteal fossa. Premature formation of the distal popliteal artery and retardation of varying degrees of the pathway of migration of the medial head of gastrocnemius results in the first two types of entrapment (Table 1).

Unlike anatomical popliteal entrapment, functional popliteal entrapment (type 6) occurs in patients with unremarkable popliteal anatomy. This is associated with gastrocnemius hypertrophy as seen in athletes and patients using anabolic steroids which can cause intermittent compression of the popliteal artery on plantar flexion.

Diagnosis and investigations
Patients present with claudication, calf swelling and paraesthesia. Active knee hyperextension followed by plantar flexion may or may not attenuate foot pulses and is not a reliable diagnostic test. On duplex, decreased peak systolic activity on active plantar flexion against resistance or knee flexion to 15 degrees is considered diagnostic. However popliteal artery occlusion on stress manoeuvres is reported in the order of 59-85% of asymptomatic patients, indicating a significant false positive rate with duplex.

Angiography is the classical investigation for PAES. Popliteal artery compression under plantar flexion at angiography strongly indicates PAES. It may demonstrate medial deviation of the artery in 24% of all cases, indicative of type 1 PAES. Alternatively it may demonstrate aneurysm or ectasia in 9%, and occlusion in 36% on dynamic manoeuvres, or irregularity of wall in the presence of normal arterial vasculature. Angiography allows assessment of the distal vasculature for embolisation but cannot differentiate atherosclerosis from

<table>
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<td>6</td>
<td>Functional entrapment of popliteal artery. No aberrant anatomy</td>
</tr>
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</table>

MHG: medial head of gastrocnemius.
anatomical entrapment in patients with occlusion. Computed tomographic angiography (CTA) (Figs. 2 and 3) and magnetic resonance angiography (MRA) can demonstrate popliteal artery and musculotendinous anatomy, and identify other pathology, e.g. adventitial cysts that mimic PAES. MRA, though expensive, is equivalent to digital subtraction angiography, and can clearly distinguish functional and anatomical PAES.

Management
Definitive management requires surgery to release extrinsic compression and preserve or restore arterial flow. If arterial damage is minimal, myotomy of the medial head of gastrocnemius or any abnormal musculotendinous slips may suffice. More severe cases require arterial repair. Interposition vein grafting outperforms thromboendarterectomy and vein patching for stenoses with a complication rate of 16.7% compared to 45.5% (p < 0.01). Bypass using vein graft remains the treatment of choice for occlusion. Patency rates have been reported at to 57–65% over a period of 8–10 years, though studies remain limited because of small sample sizes. The posterior approach is preferable for short occlusions, providing excellent visual anatomy, access to the short saphenous vein (for interposition grafting) and better cosmesis. Longer occlusions are approached medially, as they necessitate a longer bypass graft. Surgery is recommended in all patients with anatomical PAES and symptomatic cases of functional entrapment syndrome. Surgery in asymptomatic cases of functional popliteal entrapment is not justified as these patients are not at an increased risk of vascular complications.

Coeliac artery compression syndrome
Coeliac artery compression syndrome (CACS) results from extrinsic compression by the median arcuate ligament or ganglionic tissue of the coeliac plexus. Aberrant anatomy, e.g. a high coeliac origin or low diaphragmatic crus may contribute. Some dispute the existence of CACS suggesting that collaterals should prevent ischaemic symptoms and positive surgical outcomes result from coeliac plexus transection, not coeliac artery decompression. Asymptomatic coeliac artery compression exists in 3–10% of patients undergoing liver transplantation. The diagnosis of coeliac artery entrapment syndrome is made on the combined basis of clinical and radiological findings.
Diagnosis and investigations
Symptoms include weight loss, nausea, vomiting, diarrhoea and postprandial pain, usually epigastric. Some have an expiratory systolic bruit. Athletes report exercise-related symptoms. Occasionally, aneurysms cause coeliac plexus irritation, mimicking CACS. Mitral valve prolapse and pancreatice-duodenal aneurysms have also been reported in association with CACS. Mesenteric duplex and pancreatico-duodenal aneurysms have also been reported in association with CACS. Mesenteric duplex scanning is the initial investigation of choice, followed by CTA or MRA. Biplane angiography can highlight worsening coeliac artery compression on inspiration. Gastric tonometry may reveal gastric ischaemia and has been advocated as exercise precipitates ischaemia in 86% of patients with known splanchnic stenosis. Tonometry normalised in all patients who were asymptomatic following surgery, compared to only 25% of patients who remained symptomatic despite intervention.

Management
Transection of the median arcuate ligament and diaphragmatic crus proximal to the artery followed by neurolysis and transection of the ganglionic tissue relieves symptoms. Additional coeliac artery dilatation or interposition grafting improves outcomes, suggesting that decompression alone is not always sufficient. Laparoscopic decompression achieves good results when combined with endovascular methods to dilate the artery. Endovascular stenting without surgical decompression has limited long term benefit because of continual extrinsic compression resulting in re-stenosis and fracture. Data remains limited because of the rarity of the condition.

Nutcracker syndrome
This describes left renal vein entrapment between the SMA and aorta. The renal vein can be compressed if the SMA emerges more acutely, from a lower or lateral aortic origin, or descends caudally after a shorter than normal distance of 4–5 mm. Rarely the left renal vein passes behind the aorta where it is compressed against vertebrae, causing posterior nutcracker syndrome. Anterior and posterior nutcracker syndrome can occur simultaneously.

Clinical features
Renal vein compression causes renal hypertension, disruption of small renal veins and consequent rupture into the collecting system, manifesting as micro- or macro-haematuria. Patients present with flank or non-specific abdominal pain and nausea and vomiting. Reflux from the left renal vein to the left gonadal vein can result in testicular pain and varicoceole formation. Some develop pelvic congestion syndrome with genital, pelvic and thigh varices, dysmenorrhoea, dyspareunia, post coital ache and lower abdominal pain. Nutcracker syndrome mainly affects females in third and fourth decades. The precise incidence is unknown, though diagnostic features may be present on 72% of CT scans. Duplex sonography has a sensitivity of 78% and a specificity of 100%. It compares measurements of peak velocity and renal vein diameter at the renal hilum and where the SMA crosses the renal vein. A five-fold ratio is highly indicative of the diagnosis. CTA and MRA allow visualisation of the renal vein and SMA anatomy, and are increasingly popular imaging choices. The gold standard is retrograde venography and (cine-video angiography) with reno caval pressure gradient measurements. Cine-video angiography illustrates precisely the point of left renal vein compression and demonstrates reflux of contrast into the adrenal and gonadal veins from peri-ureteral and peri-renal venous collaterals, and pooling of contrast into the renal vein. The blood pressure gradient between the left renal vein and IVC needs to exceed 3 mmHg to make the diagnosis.

Management
Surveillance is a reasonable option in minimally symptomatic young patients. In particular, this is applicable to pubertal patients because of a higher rate of spontaneous remission, possibly due to physical development. Surgical options include transposition of the renal vein further down the IVC or the SMA caudally on the aorta, though this has a higher risk of complications. Auto transplantation of the affected kidney into the iliac fossa may achieve good results. Gonadal caval bypass has been described for cases with extensive pelvic varices, but the results are not encouraging. Intravascular stents have achieved good initial results, though long term follow-up is required. Management of the nutcracker syndrome may resolve haematuria, though pressure measurements may fail to reflect this improvement.

May–Thurner syndrome
Compression of the left common iliac vein between the right common iliac artery anteriorly, and the fifth lumbar vertebra posteriorly can cause stasis and lower limb deep vein thrombosis, accounting for the increased incidence of left sided DVT. May and Thurner analyzed autopsy specimens and found ‘intraluminal-spurs’ at the junction of the left common iliac vein (LCIV) with the IVC in 22% of subjects. They proposed that these were predisposing factors for ileo-femoral DVT and arose from chronic intimal damage of the LCIV secondary to pulsatile movement of the right common iliac artery. Absence of spurs in fetal autopsies suggests they are acquired, not congenital. Cockett and Thomas correlated the clinical features of this condition with its pathological findings, calling it ‘iliac compression syndrome’. Eponymous names are May–Thurner or Cockett’s Syndrome.

Clinical features
It typically affects young and middle aged women and may contribute in 18–49% of patients with left sided DVT. Symptoms include lower limb swelling and pain in the acute stage, and varicose veins, venous eczema, hyperpigmentation, exertional pain and venous ulcers in the chronic stage (post-thrombotic limb). Conventional venography delineates pelvic vein anatomy, collateral formation and iliac vein compression with consequent spur formation (Fig. 2). May and Thurner advised the use of iliac vein pressure differences (2 mm and rest and 3 mm on exercise) as diagnostic criteria. MR Venography (MRV) is the preferred imaging modality, as apart from its diagnostic
artery compressing D3. Thin body habitus may predis-

caudal pull on the mesentery during ileo-anal anasto-

thetic dilation of the duodenum without proximal stenosis

megaduodenum, though the latter differs by character-

of the third part of the duodenum (D3) secondary to

Superior mesenteric artery syndrome

Superior mesenteric artery syndrome (SMAS) is obstruction of the third part of the duodenum (D3) secondary to external compression by the SMA. First described by Von Rotikansky in 1842,53 it is also referred to as Wilkie syndrome,85 cast syndrome or arterio-mesenteric duodenal compression.86 SMAS may be confused with megaduodenum, though the latter differs by characteristic dilation of the duodenum without proximal stenosis and is secondary to cases of intestinal myopathy. Barium studies demonstrate SMAS in 0.13–0.3% of the pop-

angle.95 CTA or MRA may identify a reduced aorto-mesen-

pathology.81

Management

Anticoagulation alone is insufficient, failing to relieve the underlying mechanical compression. Compression bandages or stockings have benefits but are unpopular. Multiple surgical approaches have been described including thrombectomy in the acute stage.82 Vein patch angioplasty with intra-luminal band excision and/or interposition of a fascia lata sling or a peritoneal flap can be performed to avoid recurrent damage. The right common iliac artery can be relocated behind the LCIV or IVC. Finally a Palma Crossover procedure can be per-

Success remains variable, with 40–88% patency rates of the common iliac vein or venous bypass.79,82 None of the surgical options demonstrates clear superior-

Catheter-directed thrombolysis followed by angioplasty with stent placement has recently been reported.83,84 Mickley et al. placed percutaneous stents intraoperatively on the finding of venous spurs in radio-

local factors Aneurysm,98 neoplasm

Non-operative treatment has a success rate up to 90%,87 though it may cause blind loop syndrome.

Clinical features

Presenting features include postprandial discomfort, nausea, weight loss and early satiety. The pain is typically relieved in the lateral decubitus position or by bringing the knees to the chest which reduces tension on the small bowel mesentery. Barium studies demonstrate a dilated proximal duodenum terminating at the third part. Four criteria differentiate SMAS from megaduodenum: 1) dilata-

 unrelied in the lateral decubitus position or by bringing the

necessary to restore retroperitoneal tissue bulk and hence the aorto-mesenteric angle. Nutrition is provided by nasogastric feeding, or ideally by a nasoje-

re-occlusion rate in patients with stents following

and in severely burned patients (1.1%).90 Typically, it affects women and 75% are between 10–39 years of age.91

Pathophysiology

The SMA normally forms an angle of between 38 and 60 degrees at its aortic origin.85,87 The arterio-mesenteric distance ranges from 10 to 28 mm.87,92 This angle is maintained by retroperitoneal fat tissue, the left renal vein, lymphatic tissue and the pancreatic uncinate process. A decrease in this angle to 6–16 degrees or the mean arterio-mesenteric distance to 2–8 mm results in the artery compressing D3.87,92 Thin body habitus may predis-

pose to SMAS but an additional insult is required for the condition to manifest (Table 2). Catabolic and malnour-

ished states predispose to this condition by a reduction in mesenteric and retroperitoneal fat, hence the increased risk in patients with burns.90 All patients requiring pro-

longed bedrest following severe injury are at risk.93 Lengthening of the spine during scoliosis surgery,88,89 and caudal pull on the mesentery during ileo-anal anasto-

mosis94 reduces the aorto-mesenteric angle, leading to SMAS.

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Management

Medical management is the mainstay. Gastroduodenal decompression, hyperalimentation and correction of contributing conditions aim to restore retroperitoneal tissue bulk and hence the aorto-mesenteric angle. Nutrition is provided by nasogastric feeding, or ideally by a nasoje-

junal tube placed distal to the obstruction.99 Total paren-

ternal nutrition is a useful adjunct or an alternative if enteral feeding is impossible. Metoclopramide may enhance motility. Non-operative treatment has a success rate of 83%.96 Surgical care is warranted when medical management has been unsuccessful, especially in patients with chronic SMA syndrome with duodenal stasis and complicating peptic ulcer disease. Gastrojejunostomies provide gastric decompression but may not alleviate duodenal obstruction, causing persistent symptoms. Division of the ligament of Treitz with duodenal mobilisation (Strong’s procedure) has also been carried out, though it may lead to volvulus. A duodenojejunostomy remains the operation of choice to relieve the obstruction, with success rates up to 90%,87 though it may cause blind loop syndrome.
Recent innovations include laparoscopic duodenojejunos-tomy with lysis of the ligament of Treitz or infrarenal transposition of the superior mesenteric artery.

Conclusions

Vascular entrapment syndromes are rare, but cause significant symptoms interfering with the quality of life of young patients. Hence sound knowledge of these conditions is required to investigate and treat them using modern imaging and techniques. Imaging with MRI and CT in conjunction with endovascular and laparoscopic techniques enables us to offer less invasive therapy to patient with these interesting syndromes.

References

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