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Chronic intestinal pseudo-obstruction as the initial feature of pure autonomic failure

Chronic intestinal pseudo-obstruction (CIP) is a rare and highly morbid syndrome characterised by impaired gastrointestinal propulsion together with symptoms and signs of bowel obstruction in the absence of any lesions occluding the gut lumen (tumours, adhesive peritonitis, and so on).¹ CIP is thought to have two forms: myogenic and neurogenic.¹ Comorbid urinary retention may also occur.² Postural hypotension is not a feature in CIP. However, we recently had such a CIP patient with profound postural hypotension, which was detected only by a head up tilt test, and he was finally diagnosed as pure autonomic failure (PAF).

Case report

A 59 year old man gradually (over two months) developed intractable nausea and vomiting immediately after taking meals, although he did not have dysphagia. He also had abdominal distension, discomfort, and mild difficulty in defaecation. At that time, he had no features to suggest autonomic failure affecting other systems than the gut, such as postural dizziness or genitourinary dysfunction. Six months later, he was admitted to a gastrointestinal surgery hospital because of these symptoms, at which time he was emaciated. However, gastric and colonic endoscopy showed no organic lesions. As abdominal x ray findings continued to support the suspicion of intestinal obstruction, a partial colectomy (ileo-cecum and sigmoid colon) was carried out. However, the resected colon specimen showed no organic lesions such as a tumour, and his bowel symptoms persisted. In addition, after his indwelling urinary catheter was removed, the patient was unable to urinate and was taught to perform clean, intermittent self catheterisation (four times a day). The patient visited another gastroenterology hospital where he was shown to have delayed gastric transit. Based on these findings, he was diagnosed as having CIP. Treatment with prokinetic drugs such as itopride hydrochloride (a peripheral dopamine D2 receptor antagonist) and mosapride citrate (a peripheral 5-HT₄ receptor agonist) caused his vomiting and abdominal discomfort to be moderately ameliorated.

Seven months later, because he still had bowel and urinary dysfunction, he was referred to our hospital. Neurological examination was completely normal. Bedside systolic pressure fall on standing was small (16 mm Hg, from 142 to 126) with an increase in heart rate (22 beats/min, from 60 to 82), and he had no dizziness. Routine laboratory data and a cerebrospinal fluid examination were normal. Anti-Hu antibody and anti-nuclear antibodies were negative. Nerve conduction studies were normal in the upper and lower limbs. Brain magnetic resonance imaging (MRI) was normal. Although he had had no episodes of postural or postprandial dizziness or syncope during the course of his illness, a head up tilt test

(60° for 10 minutes) resulted in a systolic blood pressure fall of 53 mm Hg (from 178 to 125; normal <20) with a heart rate increase of 29 beats/min (from 51 to 80).³ Plasma noradrenaline was 44 pg/ml (normal >100) in the supine position and 83 pg/dl after the head up tilt.³ Although he had no complaints relating to his eyes, diluted noradrenaline and pilocarpine instillation showed denervation supersensitivity in the pupil. He did not have sicca symptoms. Since he had urinary retention, we undertook electromyography (EMG) cystometry, done according to the standards of the International Continence Society (Janus, Lifetec Inc, Houston, Texas, USA). He could not urinate because of an acontractile detrusor and non-relaxing sphincter. In addition, the patient had completely lost first bladder sensation (>600 ml; normal, 100 to 300). Although he reported that his erection was normal, Rigiscan Plus (Timm Medical Technologies Inc, Minnesota, USA) showed markedly decreased nocturnal penile tumescence. Under cessation of the prokinetic drugs, we also carried out gut function tests following the standards set by the American Gastroenterological Association, using electrogastrography (Nipro Inc, Osaka, Japan), a colonic transit time (CTT) test (Sitzmarks; Konsyl Pharmaceuticals Inc, Edison, New Jersey, USA), and recto-anal videomanometry (Janus, Lifetec Inc),⁴ even though he had undergone the partial colectomy. The electrogastrography did not show the postprandial or diurnal acceleration that were observed in normal subjects. The patient had a prolonged total CTT of 72.0 hours (normal 16.0 to 48.0). In the videomanometry, during rectal filling with contrast medium (50 ml/min), he did not have the phasic rectal contractions that are observed in normal subjects. All these findings confirmed the diagnosis of PAF.

Comment

Our patient had initially been diagnosed as having CIP because he had abdominal distension without any organic lesions on gastric and colonic endoscopy.¹ CIP can be classified as either secondary to a wide array of recognised pathological conditions or idiopathic. The known mechanisms of CIP include "myogenic" and "neurogenic" types.¹ In myogenic CIP, vacuolar degeneration and fibrosis of the muscularis propria occur (also known as visceral myopathy). De Giorgio *et al*¹ further classified neurogenic CIP into two forms: *inflammatory neuropathies*, in which a significant inflammatory/immune response is identified within the myenteric ganglia (anti-Hu antibody-positive CIP, vasculitis associated CIP, and so on); and *degenerative neuropathies*, characterised by evidence of neurodegenerative aspects or mitochondrial abnormalities. It is noteworthy that our CIP patient had postural hypotension suggestive of generalised autonomic failure, which was detected only by a head up tilt test, and he was finally diagnosed as having PAF. PAF is an idiopathic sporadic disorder characterised by postural hypotension, but usually with more widespread autonomic failure including bowel and bladder dysfunction.^{3–5} No other neurological features are present, such as gait difficulty or numbness, distinguishing this disorder from multiple system atrophy or amyloid neuropathy. Postural hypotension is usually the initial and presenting feature in PAF. However, when postural hypotension progresses insidiously and the systolic blood

pressure on standing is maintained above 100 mm Hg, postural hypotension might not make itself obvious to the patient. Thus postural blood pressure should be checked to determine the extent of autonomic abnormalities in CIP.

Our case suggested that testing for postural hypotension should be carried out to determine the extent of autonomic abnormalities in CIP, as PAF may present with CIP as the first or presenting symptom.

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doi: 10.1136/jnnp.2005.079905

Competing interests: None declared.

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Remission of chronic inflammatory demyelinating polyneuropathy after alemtuzumab (Campath 1H)

We describe a patient with intravenous immunoglobulin (IVIg) dependent relapsing chronic inflammatory demyelinating polyneuropathy (CIDP), unresponsive to steroids or conventional immunosuppressive agents, who achieved remission following treatment with alemtuzumab.

A 19 year old woman presented with a two week history of distal lower limb paraesthesiae and distal upper limb weakness. Her symptoms worsened over four weeks but she remained mobile. There was no history of preceding infections, pain, or autonomic dysfunction. Examination revealed reduced limb muscle tone and distal weakness, more marked in the upper limbs. She was areflexic with flexor plantar responses and had