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Severe isolated myopathic gastroparesis: a case report with pathological findings

Normal gastric emptying is a complex mechanism involving coordinated gastric and duodenal muscular contractions modulated by the autonomic nervous system, myenteric and submucosal plexus, interstitial cells of Cajal (ICC), which are divided into myenteric (ICC-MY) and intramuscular ICC (ICC-IM), and the central nervous system. Gastroparesis can result from a lesion at any level of these physiological processes.

In 1997, a 47 year old man was referred to Louis Mourier Hospital with longstanding symptoms of gastroparesis associated with pyrosis and regurgitations with progressive worsening. Physical examination and usual laboratory results were normal. An autoimmune study was negative for every antibody, including anti-HU, anti-YO, anti-RI, anti-nuclear, anti-RO, anti-SSA, anti-intrinsic factor, antigliadin, antiendomysium, anticardiolipin, antithyroxine, and antithyroglobulin antibodies. Serum antibodies of the herpes class virus, Epstein-Barr virus, and cytomegalovirus were in favour of past infections. Upper gastrointestinal endoscopy with histological analysis was normal. Gastric scintigraphy showed severe delayed gastric emptying. Antroduodenal manometry demonstrated total absence of antral motor activity, while intestinal activity was strictly normal. A brain computed tomography scan and an electromyogram of the four limbs were normal. The diagnosis of isolated severe gastroparesis was retained. Prokinetic treatments were unsuccessful. After eight months of enteral nutrition, and due to symptom severity, total gastrectomy with an oesophageal anastomosis was performed. After surgery, oral feeding was reintroduced. Nine years later the clinical condition remains satisfactory and no other motility disorder has appeared.

The gastric macroscopic appearance was normal. Histological examination revealed atrophic gastritis with intestinal metaplasia in the antral and fundic areas, with a *Helicobacter pylori* related active interstitial gastritis. The submucosa was normal. In the antrum, the inner circular muscle layer was normal while the outer longitudinal layer was missing or reduced to a few fibres (fig 1). In

Figure 1 Antrum: the outer layer of the muscularis propria was reduced to a thin layer (arrow) or had disappeared completely (*). Haematein eosin saffron, original magnification $\times 25$.

Figure 2 Intermuscular plexus: ganglion cells were normal. Note the inflammatory infiltrate consisting of a few CD8 positive T cells. (A) Haematein eosin saffron; (B) Immunohistochemistry, anti-CD8, DAB peroxidase; original magnification $\times 250$. (C) Note the absence of Cajal cells. Mast cells were positive controls (arrows). Immunohistochemistry, anti-CD117, DAB peroxidase; original magnification $\times 250$.

some myenteric plexus, an inflammatory infiltrate was present, consisting of small regular CD3⁺, CD8⁺, CD4⁺, and CD20⁺ lymphocytes (fig 2). In the antrum, ICC (CD117⁺) and their extensions were lacking or rarefied in the myenteric plexus (fig 2) whereas ICC-IM had a normal distribution. Ultrastructural examination showed no abnormality in the inner circular muscle layer. ICC were visualised in small numbers but appeared normal.

Our final diagnosis was myopathic gastroparesis which has not been reported previously in adulthood. In the antrum, the primary component of each slow wave is initiated by ICC-MY and increased by the secondary component initiated by ICC-IM. ICC-IM are intermediary cells in the pathway by which nerves modulate the excitability of the circular muscle layer.¹ In the longitudinal layer of the antrum, ICC-IM are normally present in very low numbers.¹ Presumably, neurally released transmitters can directly access smooth muscle cell membranes. In the case of the lesion of the ICC-MY, as in our patient, we can hypothesise that the outer longitudinal muscular layer could be more sensitive than the inner circular layer and could become atrophied. In a murine model, gastric smooth muscle atrophy can lead to ICC depletion due to a deficit in stem cell factor (SCF) production.² Enteric ganglionitis can be primitive or secondary to a wide array of diseases. In our patient, the longstanding and progressive evolution of symptoms as well as pathological lesions were not in accordance with infectious gastroparesis,³ and the symptomless nine year follow up period excludes any paraneoplastic syndrome.⁴ Finally, in our patient, inflammatory lesions were unusual as only part of the

myenteric plexus was involved and was exclusively located in the antrum.

Taken together, these data suggest that, in common with intestinal chronic pseudo-obstruction, severe idiopathic isolated gastroparesis may be related to myopathic as well as neuropathic disorders.⁵ As has been suggested in chronic intestinal pseudo-obstruction and inflammatory enteric neuropathies,⁴ full thickness gastric biopsy performed under laparoscopic control may be helpful in the management of these severely affected patients.

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An exuberant inflammatory response to *E coli*: implications for the pathogenesis of ulcerative colitis and pyoderma gangrenosum

A recent study investigated acute inflammatory responses in patients with inflammatory bowel disease.¹ Subjects received 1 mg of heat killed *Escherichia coli* subcutaneously into each forearm. The magnitude of the ensuing local inflammatory responses, measured as increases in local blood flow, peaked in healthy individuals by 24 hours and resolved within 48 hours. In contrast, the response in two patients with ulcerative colitis was abnormally protracted (supplementary fig 1; supplementary fig 1 can be viewed on the Gut website at <http://www.gutjnl.com/supplemental>).

One of these patients, a 24 year old female with a 10 year history of left sided ulcerative colitis (confirmed histologically and quiescent on olsalazine 2 g daily), developed severe local complications at both injection sites. Her initial local reaction was macroscopically similar to healthy controls but persisted, progressing to painful erythematous swellings (fig 1A) with vesicobullous