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Editorial

Intestinal pseudo-obstruction: a diagnostic and therapeutic challenge for the 21st century

Since Dudley's introduction of the term intestinal pseudo-obstruction back in 1958 (1), many cases and patient series have been reported in the literature, and various classifications have been suggested; however, few large studies managed to unearth all the mysteries underlying what we have come to know as a chronic intestinal pseudo-obstruction (CIPO). Only in the past few years patient series large enough to allow conclusions were published (2,3).

In this issue of *Revista Española de Enfermedades Digestivas,* Muñoz-Yagüe et al. (4) provide us with a singular case of CIPO that might be classified as a non-familiar visceral myopathy with mainly colonic involvement. The exemplary study of the patient reveals the profound clinical orientation of the author's school of thought, with a vocation to concurrently solve the patient's ailments and reach the core of the matter.

The case has a number of singular aspects. First, primary CIPO in the adult may reflect a neuropathy, mesenchymal disease (interstitial Cajal cell involvement) or myopathy (5,6). While we lack reliable data thereupon, the former condition seems more common (3,7), even if that is not everybody's experience (8). Second, involvement is mainly colonic in the reported case, and the small bowel was seemingly little affected, which is unusual for a CIPO description, even though said affectation existed at the histological level with no apparent clinical expression; intestinal manometry, while not strictly necessary in the diagnosis of CIPO (3), might have helped reveal whether such histologic involvement had a correlate in the patient. This predominantly colonic involvement, and its histologic descriptions are all similar to definitions regarding degenerative leiomyopathy (9), initially described as Bantu's pseudo- Hirschsprung's disease, a non-familiar visceral myopathy seen in African children exclusively. Because of its current relevance, the use of alpha actin expression, recently suggested for the diagnosis, deserves mention; the patient had a deficient expression in the small bowel and colon, a finding considered a marker (7), not a cause, whose relevance is not supported by all (10) and not considered by some to be myopathy-specific (11). Finally, the patient's outcome singularity cannot be overstated in that it was relatively benign, which is not common in CIPO; the development of proximal tetraparesia leads to consider a potential mitochondrial myopathy (6), which the authors rule out.

The case reported by Muñoz-Yagüe et al. should prompt reflection on the need to keep senses alert regarding CIPO, and good clinical judgement may provide us with an understanding of our patient's difficulties, leading to the delivery of the best diagnostic and therapeutic services available. On the other hand, one should wonder whether relatively rare conditions such as CIPO deserve a concentration of effort not only with the objetive of achieving better results, but also to increase our understanding and respond to the trust of future patients in 21st century medicine.

Editorial

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