PICTURES IN DIGESTIVE PATHOLOGY

Hirschsprung's disease in adults

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Fig. 1. Computed tomography, where the sigmoid colon (9 x 11 cm diameter in axial cuts) is seen, with faeces inside, which displace the abdominal viscera.

Fig. 2. Barium enema with estenotic area in the upper rectum, marked with the arrow, and compensative dilatation of sigmoid colon.

CASE REPORT

A 19-year-old man came to the emergency room because of diarrhea of 4 weeks duration, accompanied by constipation. As background, he reported fecal incontinence with encopresis since he was 12 year-old, initially ascribed to a child psychiatric disorder. Physical examination revealed an important thinness (BMI 18,51 kg/m²) and a large mass in the left abdomen, of hard consistency, mobile, and painless. Abdomen X-ray examination showed large dilatation of descending colon, with distal air. Computed tomography (Fig. 1) disclosed a highly dilated sigmoid colon containing feces inside; the distal portion of the rectum was normal. These radiological images were consistent with Hirschsprung's disease; for this reason, we extracted surgically the fecal mass through the anus. After that, the palpable mass disappeared, but clinical constipation persisted. Later, a barium enema (Fig. 2) was carried out and results were compatible with that disease.

Therefore, a full thickness rectal biopsy was performed, where ganglion cells were absent (Fig. 3), what confirmed the diagnosis. He is awaiting surgical intervention.

DISCUSSION

Some authors (1-3) have maintained that Hirschsprung's disease in adults probably correspond to congenital cases not diagnosed previously, and consequently it is likely that, in fact, adult Hirschsprung's disease does not really exist as an independent entity.

Although, this is an uncommon cause of chronic constipation in adults (4), it should be considered in the differential diagnosis of this entity in similar clinical cases, as the herein presented. Undoubtedly, it represents a diagnostic and therapeutic challenge.

Fig. 3. Depth rectal biopsy, neuron-specific tinction (10x) showing the lack of ganglion cells. These characteristics define this entity.

REFERENCES

1. Haricharan RN, Georgeson KE. Hirschsprung disease. *Semin Pediatr Surg.* 2008;17:266-75.
2. Chen F, Winston III JH, Jain SK, Frankel WL. Hirschsprung's disease in a young adult: report of a case and review of the literature. *Ann Diagn Pathol* 2006;10:347-51.
3. Chatelain D, Manaouil C, Marc B, Ricard J, Brevet M, Montpellier D, et al. Adult Hirschsprung's disease diagnosed during forensic autopsy. *J Forensic Sci* 2006;51:1160-3.
4. Yüksel I, Ataseven I, Erturk I, Başar O, Akmaz N. Adult segmental Hirschsprung disease. *South Med J* 2009;102:184-5.