

Case	(023) Adult hirschsprung's disease
Authors	P. Marazuela García, J.v. Quintana Pérez, B. Alba Pérez, A. López-frías Lópezjurado, J. Blanc Molina, E. García Casado.
Centre	Hospital Universitario Ramón Y Cajal.

CASE PRESENTATION

33 year old woman attending the emergency room because of one week abdominal distension and moderate efforts dyspnea. Patient also refers constipation of at least three months of evolution, which has managed with laxatives and enemas. No fever, nausea neither vomits, no abdominal pain and no other significant symptoms. No other personal background of interest.

In the laboratory results, highlights exclusively low albumin (3,28 g/dL).

IMAGE FINDINGS:

A, B: In the chest and abdominal X-rays we identify severe dilatation of the colon with important amount of feces. It is not clear which segments are more pathologic or if there is pneumoperitoneum.

C, D: IVC abdominal CT shows dilatation of the sigma until 24 cm. The other colonic segments present normal caliber except for the hepatic angle, which is slightly expanded. This is because of the back compression of the transverse colon. All the other abdominal organs, and even the lungs and the heart, are also compressed and displaced. In image D we identify a short segment of the recto-sigmoid union collapsed, which could be the aganglionic segment. No volvulus signs are identified.

DISCUSSION

Although very rare, HD can be newly diagnosed in adulthood. Patients present abdominal distension and a long history of refractory constipation without fecal incontinence.

Abdominal X-Ray shows massive dilatation of proximal colon to the aganglionic segment. CT should be performed in order to exclude other causes of megacolon, such as volvulus or colorectal cancer. Volvulus could also be an associated complication of adult HD, presenting more acute signs of intestinal obstruction or even progress to bowel ischemia.

The presence of a "transition zone", representing the change from the normal caliber/narrowed rectum (aganglionic segment) to the dilated colon proximal to the aganglionic region, is virtually pathognomonic of HD. This finding is usually located in the rectosigmoid area, and despite being a concept employed in barium enema, it can also be seen in CT.

"Ultra-short" segment Hirschsprung's disease is sometimes used to describe a form characterized by a very short segment of aganglionosis extending from 2 to 4 cm proximal to the internal anal sphincter.

CONCLUSION

Hirschsprung's Disease is typically diagnosed in neonatal period, however it can cause megacolon in adults with less severe disease. CT scan in the emergency room should be performed in order to dismiss other motives of megacolon, identify possible volvulus and sometimes determine the transition zone.



BIBLIOGRAPHY

-López Ruiz JA, Tallón Aguilar L, Sánchez Moreno L, López Pérez J, Pareja Ciuró F, Oliva Mompeán F, et al. Hirschsprung disease with debut in adult age as acute intestinal obstruction: case report. *Revista Española de Enfermedades Digestivas*. 2016;108. Available in: <https://online.reed.es/fichaArticulo.aspx?iarf=224686761-749235413274>

- N.L. Crocker and J.M. Messmer. Adult Hirschsprung's disease. *Clinical Radiology*. (1991) 44, 257-259.