Intestinal pseudo-obstruction is related with enteric nervous system disorders. Intestinal neuronal dysplasia is an aganglionosis and is different from Hirschprung’s disease as it is a hyperplasia of the submucosal and myenteric plexuses, with ganglion hypertrophy. We present the case of a 47-year-old patient, with symptoms of progressive respiratory failure and severe abdominal distension due to massive megacolon and diaphragm compression (Fig. 1). After a failed colonoscopy, a laparotomy and a decompression colostomy were performed. Two months later, a subtotal colectomy with ileorectal anastomosis was done (Fig. 2), and the patient’s evolution was favorable. The pathology study diagnosed intestinal neuronal dysplasia.

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