Clinical picture

Acute urinary retention from coprostatic colon: an unusual presentation of ‘adult Hirschsprung’s disease’

Introduction
Hirschsprung disease (HD), or congenital intestinal aganglionosis, is characterized by absence of neurons in the distal colon resulting from a lack of migration of neural crest cells to the large intestine during fetal development.1 HD typically manifests in early childhood, but very rarely the diagnosis may be delayed for several years when ‘adult HD’ patients present with refractory constipation, intestinal obstruction, sigmoid volvulus or colonic perforation.2 While co-existing congenital genitourinary anamolies have been reported in 6% of HD cases, the extra-intestinal manifestations such as hydronephrosis and urinary retention are extremely rare.3–5 Here, we present first reported case to our knowledge of an adult HD patient presenting with urinary retention from an extrinsic compression of the urinary bladder by a distended ‘coprostatic’ colon.

Case
A 21-year-old male presented with 1 week of worsening diffuse abdominal pain and distention without any bowel movement. This was associated with an acute difficulty in voiding urine. He had suffered from constipation since early childhood and was diagnosed with HD on a rectal biopsy about 3 years ago, but failed to follow-up for surgical management. Severe abdominal distention and tenderness at this presentation prompted diagnostic testing with a contrast enhanced computed tomography (CT) scan. The distal colon and recto-sigmoid were markedly dilated with impacted stool, and measured 11 and 9 cm in diameter, respectively, whereas the proximal colon measured 5 cm (Figure 1). The bladder was massively distended and displaced into the left side of the abdomen, without evidence of hydronephrosis. Renal function tests were within normal range. Approximately 1000 ml of urine was drained at the time of a Foley’s catheter placement indicating urinary retention. He underwent an urgent sigmoid loop colostomy to decompress his feces laden colon. In the post-operative period, he had gradual improvement of his stomal output and regained normal bladder function prior to discharge. He was recommended definitive surgery for repair of his aganglionic colon.

Discussion
Adult HD is a very rare condition posing a diagnostic and therapeutic challenge. Chronic refractory constipation in childhood and adolescence should be thoroughly investigated to diagnose HD. Uncommon urinary complications such as urinary retention have to be borne in mind in the appropriate clinical setting. Prompt diagnosis and surgical repair of the aganglionic segment is crucial for restoring colonic function and avert life-threatening complications.
Photographs and text from: P. Loganathan; K. Vipperla, C. Umapathy and M. Gajendran, Division of Gastroenterology, Hepatology and Nutrition, University of Pittsburgh, M2, C Wing, 200 Lothrop Street, Pittsburgh, PA 15213, USA and Division of General Internal Medicine, University of Pittsburgh, Suite 933W, 200 Lothrop Street, Pittsburgh, PA 15213, USA. 
email: gajendranm@upmc.edu

References