Re: Kumar M, Batra G, Maletha M, Malhotra A. Hirschsprung's disease in a child with posterior urethral valve: An unwanted association. Indian J Urol 2018;34:223-5

Dear Sir,

We read the article titled "Hirschsprung's disease (HD) in a child with posterior urethral valve: An unwanted association" by Kumar *et al*.[1] with great interest, and we congratulate the authors for successfully managing and reporting this rare coexistence of a posterior urethral valve and bowel dysfunction in a 20-months-old child. We offer the following comments.

The authors have shown dilated regions of the rectum and proximal sigmoid with a focal area of narrowing in between the two segments in the contrast enema film. This particular appearance on a barium enema is pathognomonic of congenital colonic stenosis or a postnecrotising enterocolitis (NEC)-related stricture of the colon, rather than HD. There is enough evidence that neural crest cells migrate in a craniocaudal direction. Failure of their distal migration and inability to survive (and differentiate) in the distal gut are possible theories of the etiopathogenesis of HD.[2] This produces a spastic distal aganglionic colon which is the hallmark of HD of the rectosigmoid region in 80% of the cases.[3] Therefore, an isolated (focal) region of narrowing with a distensible distal rectum elaborates a possible scenario of congenital colonic stenosis or an NEC-associated stricture, rather than HD.

The lack of ganglion cells reported as "rectal biopsy confirmed the absence of ganglion cells in the affected segment of the sigmoid colon" is confusing and needs to be further refined to specify the exact site of the biopsy. Assuming it to be a rectal biopsy, we may not forget that making a false-positive histopathological diagnosis of HD using mucosal biopsies or biopsy from the terminal 1–3 cm (physiological zone of aganglionosis) is not uncommon.[4] With these concepts, it must be evident that the diagnosis of "ultrashort-segment" HD is rather difficult to be made on barium enema (indiscernible or no transition zone) and also by rectal biopsy (high false negative). Thus, in a suspected case of "ultrashort-segment" HD, anorectal manometry may have helped in clinching the diagnosis.[5]

It will also be of interest to the readers, if the author clarifies the operative steps, since in cases of "ultrashort-segment" HD, a myectomy only may suffice.[6] It gives an impression from the report that the authors have performed a resection of the focal area and anastomosis which is beyond the principles of surgical management of HD or "ultrashort-segment" HD. This clarification becomes all the more relevant since there is a controversy in the literature regarding the diagnosis and management of "ultrashort-segment" variant of HD in contrast to the "rectosigmoid" form of HD, where the diagnosis and management are standardized.

We hope that these points enable the authors to reconsider their diagnosis of "ultrashort" HD in this case.

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