TOTAL COLONIC AGANGLIONOSIS (ZUELZER WILSON SYNDROME) An Enigma

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Introduction

Most children of total colonic aganglionosis (TCA) present with distal intestinal obstruction in the neonatal period. However, in some children it is relatively common to pass some stools and not be instantly recognised as having aganglionosis with distal ileal involvement presenting late in life. Appropriate surgical management of TCA, which can often be a diagnostic dilemma, is discussed.

Case Report

A one year old boy was admitted with complaints of chronic constipation with history of repeated diarrhoea and vomiting. Mother also gave history of delayed passage of meconium after birth. Examination revealed a poorly nourished child with height of 71 cms (ie. less than 10th percentile curve) and weighing 6 Kg (i.e. below the 3rd percentile of body weight). There was distension of abdomen and per rectal digital examination was followed by passage of copious amount of foul smelling faeces. While in the hospital the baby developed enterocolitis in the form of foul smelling loose stools, vomiting and fever. He was treated with IV

fluid, broad-spectrum antibiotics and rectal washouts to which he responded well. A clinical diagnosis of the Hirschsprung's disease was entertained.

Barium enema did not reveal any transitional zone or micro-colon. Rectal biopsy confirmed rectal aganglionosis. Exploratory laparotomy revealed a normal caliber colon with a classical transitional zone in the terminal ileum (Fig 1). There was no organic intraluminal obstruction. Multiple seromuscular biopsies were taken from the rectum upwards to transitional zone including the removal of appendix. A full thickness rectal biopsy was also taken. The ileostomy was placed just proximal to the transitional zone. Histopathological examination revealed absence of ganglion cells in all samples excepts from ileum proximal to the transitional zone thus confirming the diagnosis of total colonic aganglionosis (Fig 2).

The post ileostomy diarrhoea was troublesome and therefore definitive surgery was done 3 weeks after fashioning of the ileostomy. The surgical procedure included a total colectomy with removal of terminal ileum, ileostomy stoma and a pull through of the proximal ganglionic ileum retrorectally by the Duhamel Martin's technique. A 10 cms long segment of sigmoid colon was retained which was sutured side to side with the pulled through ileum. The intervening spur was crushed by an Ikeda clamp. No covering ileostomy was done. Post-operatively child had persist-

Fig. 1 : Operative photograph showing total colonic aganglionosis with terminal ileal involvement. A cone shaped transitional zone is clearly visible in the terminal ileum

Fig. 2 : Multiple thick nerve bundles are seen between layers of smooth muscles but no ganglion cells are seen (HE 10x)

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ent diarrhoea, as the intestinal adaptation had still not taken place. However, this improved in due course of time. The child has gained 2 Kgs in the immediate post-operative period (i.e. 3 weeks after operation) and is under close follow up.

Discussion

Total colonic aganglionosis (TCA) was first reported by Zuelzer and Wilson [1], who suggested that aganglionosis could extend from the duodenum to the rectum. Total colonic aganglionosis occurs in 5% to 8% of all patients with Hirschsprung's disease [2]. Delayed passage of meconium, constipation and abdominal distention are the most common presenting signs and symptoms of the Hirschsprung's disease. Severe iron deficiency anaemia, low levels of B12 and growth retardation may be apparent in patients with TCA with ileal involvement.

The diagnosis of total colonic aganglionosis remains a difficult problem. Diagnostic difficulties result in therapeutic delay or inappropriate surgical treatment. No radiographic finding is pathognomonic for TCA, though a colon of normal or irregular calibre with foreshortening is suggestive. More importantly, nonevacuation of colonic contrast material 24 hours after contrast enema suggests the diagnosis [3]. When suspected, rectal biopsy proves the existence of aganglionosis which is determined at laparotomy. Frozen section studies of seromuscular biopsies and of the appendix are helpful in determining the transition zone, as we did in our case.

The basic principle involved in treatment of Hirschsprung's disease is resection of aganglionic segment and restoration of intestinal continuity. However, TCA with or without small bowel involvement is difficult to manage as the aganglionic zone is long and resection results in loss of absorptive surface. The external diversion (caecostomy or ileostomy) produces hazardous results due to excessive fluid and electrolyte loss as a result of high diversion and nutritional problems due to loss of variable length of small intestine. Total colectomy is also difficult to perform in newborns as a one-stage procedure.

The first operation designed specifically for the management of total colonic aganglionosis was Lester Martin's modification of the Duhamel procedure [4]. Martin suggested that the entire aganglionic colon should be attached to the side of the normal ileum for better absorption. The initial results were encouraging but later on problems of life threatening enterocolitis and obstruction were realized due to inefficient emptying of the long aganglionic segment. In subsequent modifications Kimura and co-workers described a technique for doing a Swenson's type of operation

utilizing right half of the colon [5]. Shandling described the use of an extended ganglionic - aganglionic common channel with a Soave anastomosis [6]. Scott Boley used ileocolostomy from right colon and terminal ileum as Soave's endorectal pull through to improve functional outcome [7].

Although all procedures are reportedly successful, all have limitations and none has been proved superior. The long-term outcome of these patients revealed that the use of extensive lengths of aganglionic bowel to maximize fluid absorption was associated with substantial morbidity. Hence, Shermata and Meller by using right colon recommended that a 10 cm ileocolostomy is appropriate to control ileostomy diarrhoea [8]. We also constructed the 10 cms ileocolostomy using sigmoid colon as described by Martin.

The prognosis of the ailment is very poor with a mortality of 50%. Maximum deaths occur during the interval between the two procedures, primarily related to ileostomy complications. Enterostomy alone carries a mortality rate of more than 60% [9]. Recently Hoehner et al [10] claim improved results with 97% survival in this disease. The significant improvement in survival is a result of early recognition and diagnosis, progress in parenteral and enteral nutrition with improvement in supportive care and innovation of various surgical techniques.

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