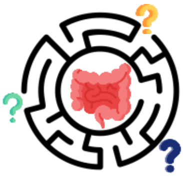

A COLORECTAL ENIGMA: RARE COMBINATION OF ANAL STENOSIS, HIRSCHSPRUNG DISEASE AND PRESACRAL TAILGUT CYST

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INTRODUCTION:

- Association between anorectal malformation and Hirschsprung disease (HD) is rare; anal stenosis (AS) has incidence <1%, with excellent functional prognosis once adequate calibre is ensured;
- Our case: bowel obstruction in which AS with presacral mass initially hindered HD diagnosis.

CASE PRESENTATION:

1 FIRST PRESENTATION: 20-day-old male with bowel obstruction and failure to thrive:

- Born at term, weight 3365 g, normal emission of meconium;
- Evaluation of perineum: AS, anal dilation (3→9mm Hegar); laparotomy: homogeneous bowel dilation, no mechanical obstructions;
- MRI: cystic presacral mass.

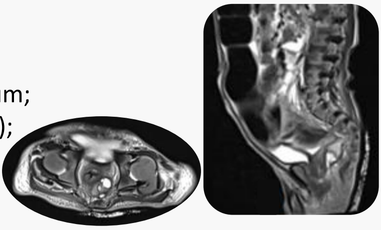

Fig 1: cystic presacral mass.

2 SECOND PRESENTATION with persistent, severe bowel obstruction (2 months, 3320 g):

- Evaluation of perineum: persistent AS (dilated to 10mm Hegar); laparotomy: transition zone at proximal sigmoid, thus colonic mapping and ileostomy;
- Histology: HD up to middle sigmoid.

3 RECONSTRUCTIVE SURGERY for classical HD (4 months, 4200 g):

- Laparotomy with complete aganglionic tract mobilization, en bloc resection of presacral mass, coccygectomy;
- Perineal stage: intussusception of aganglionic tract into the anus, extracorporeal Swenson dissection with coloanal anastomosis.
- Histology: ganglia on descending colon, tailgut cyst for presacral mass.

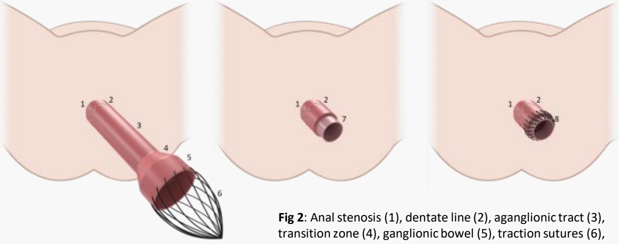

Fig 2: Anal stenosis (1), dentate line (2), aganglionic tract (3), transition zone (4), ganglionic bowel (5), traction sutures (6), ganglionic descending colon (7), coloanal anastomosis (8).

4 RECONSTRUCTIVE SURGERY for AS (8 months):

- Heinecke-Mikulicz strictureplasty obtaining stable 14mm Ø;
- Then, stoma reversal.

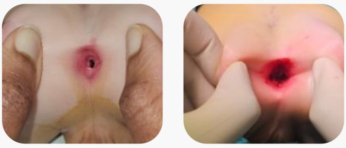

Fig 3: pre-(left) and after-anal strictureplasty (right).

5 FOLLOW-UP:

- Intrasphincteric botulinum injection for mild obstructive symptoms;
- Currently: regular bowel actions and appropriate weight gain.

CONCLUSION:

- Managing combined colorectal anomalies is complex and requires high colorectal expertise;
- A stepwise, individualized approach guided by detailed anatomical assessment is critical in planning and executing appropriate surgeries aiming at optimal long-term outcomes.

REFERENCES:

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