Surgical conservative treatment for Bauhin's syndrome

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Abstract

Background and study aims: *Hypertrophy of ileocaecal sphincter seems to be the basic etiological factor of Bauhin's valve syndrome (BVS). In the rare literature all cases are treated by means of an hemicolectomy. A patient with Bauhin's valve syndrome is described, whose pathologic characteristics were hypertrophy of ileocaecal sphincter and a circular submucosal lipoma on the caecal side of the valve.*
Lipomata, although uncommon, may arise throughout the whole gastrointestinal tract, mostly asymptomatic, and submucosal layer is most frequently involved than subserosal one. More than two-thirds of gut lipoma are found in the large bowel, where they represent the most common benign lesion after adenoma.
Patients and methods: *A 36 year old woman suffering from periodic upper abdominal pain, nausea and diarrhea, was bubmitted to an explorative surgical procedure, after imaging study of the bowel showed only an indistict mass in the caecum. Though a caecotomy a dissection of an hypertrophic and swollen portion of the ileocaecal valve was performed, saving the ileo-caecal sphincter. The residual mucosal margins were sutured, the last tract of the ileum was fixed to the caecum, completing with a caecorrhaphy and appendectomy.*
Results: *Normal post-operative period, discharging the patient after few days. Since the operation all symptoms disappeared, and after four years there is a total well-being of the patient with complete disappearance of the former symptoms.*
Conclusions: *If the BVS is only due to a lipomatous hypertrophy of the mucosa and submucosa of ileo-caecal valve, hemicholectomy seems to be not justified: it is an exceedingly invasive procedure for a non-malignant disease.*
It is important a throughout radiological diagnosis and a caecotomy direct control during an operative exploration for a correct surgical choice.
Key words: Ileocecal valve, lipohyperplasia, imaging diagnosis, sphincter hyperactivity, Bauhin's syndrome.

Riassunto

TRATTAMENTO CHIRURGICO CONSERVATIVO DELLA SINDROME DI BAUHINO

Premessa: *L'ipertrofia dello sfintere ileo-ciecale sembra essere la causa etiologica di base della sindrome della valvola*

Introduction

The history of BVS is traced by few phases. In 1953 Debray et al. described *la Bauhinite Edèmateuse: edème inflammatoire pseudo-tumoral de la valvola de Bauhin* (1). Such syndrome was characterized by recurrent diarrhea, weight loss and x-ray pictures showing enlargement of the ileocaecal valve. Histologic examination of ileocaecal junction revealed aspecific edema of the valve. Later similar cases were reported by Davidovitch (1954), Sauer, Hodgson, Mayne, and Judd (1955), Baumel (1958), and Davouze (1958) (2, 3).
Lasser and Rigler (1955) and Grayson (1958) reported a number of cases where an enlargement of Bauhin's valve was observed on x-rays. These patients had abdominal swelling and pain mainly located at the right regions, and a hypersensitivity at palpation of the right iliac fossa. These authors named this clinical and x-ray picture *"ileocaecal valve syndrome"* (4, 5).
In 1964 Gazet studied the syndrome from a pathophysiological point of view. He performed hemicolectomy in a patient suffering from ileocaecal valve syndrome. Histologic examination revealed submucosal lipoma involving ileocaecal juncture. Extensive hemorrhage areas, macrophages containing hemosiderin and fibrous bands alternated with lipophagic reaction appeared between the lipoma and the overlying muscolaris mucosae. Adipose tissue was accurately removed, noting that it did not

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di Bauhino (SVB). In letteratura tutti i casi sono trattati con emicolectomia destra. L'osservazione personale riguarda una paziente con SVB da ipertrofia dello sfintere ileo-ciecale associata ad un lipoma sottomucoso del versante ciecale della valvola.
I rari lipomi del tratto gastroenterico possono insorgere ovunque, interessando più spesso la tonaca sottomucosa, per lo più asintomatici. Più di due terzi di essi si localizzano nel grosso intestino, dove rappresentano la lesione benigna più comune dopo gli adenomi
Osservazione personale: *Donna di 36 anni, sofferente per ciclici dolori addominali con nausea e diarrea, sottoposta a laparotomia esplorativa dopo studio completo per imaging dell'intestino, per la presenza di una massa indistinta a livello ciecale. Mediante ciecotomia venne dissecata la porzione ipertrofica e sporgente della valvola ileo-ciecale, preservando lo sfintere. I due residui margini mucosi vennero suturati, l'ultima ansa ileale fissata al cieco, e dopo ciecorrafia in doppio strato, appendicectomia.*
Risultato: *Periodo postoperatoria normale, con dimissioni dopo pochi giorni. Fin dal postoperatorio tutti i sintomi scomparvero, e dopo quattro anni la paziente riferisce totale benessere e scomparsa dei sintomi precedenti.*
Conclusioni: *Se la SVB è dovuta unicamente ad un ipertrofia lipomatosa della sottomucosa della valvola, l'emicolectomia non sembra giustificata, perché troppo invasiva per una patologia non maligna.*
È importante la diagnostica radiologica preoperatoria e il controllo diretto della lesione durante l'esplorazione chirurgica per poter procedere ad una corretta scelta operativa.

invade neither the ileocaecal sphincter nor terminal ileum. Then, Gazet cut 0.5-1.5 cm long strips of circular muscle near the junction, and studied their contractility by putting them in a solution containing acetylcholine, adrenaline, noradrenaline and phentolamine. The results showed a quantitative response which was double or triple that of a normal sphincter in relation to the dose of drugs used. Furthermore, the sphincter was 2 cm long instead of the "normal" 0.5-1 cm (1).
The clinical characteristics of the syndrome are represented by symptoms which are constant: recurrent diarrhea, abdominal swelling and pain involving the right regions of the abdomen, mostly in the iliac fossa, and rare bleeding (6, 7). Usually the disease involves young adults with a ratio female/male of 2:1; it is rare in children and in over 60's (8, 9).
More frequently the condition is named "chronic colitis" and it is as such unsuccessfully treated.
Etiology is variegated due to the different experiences and opinions. It has been observed that Bauhin's valve lesion associated with the syndrome is not necessarily an aspecific edema (10, 11) as suggested by Debray et al. (1953).
Bauhin's valve syndrome (BVS) is very rare. Since its first description by Debray et al. in 1953, only 15 cases have been reported.The most recent one was described in 1995. To date, right hemicholectomy has represented the most common surgical treatment. We have recently diagnosed BVS with modern imaging means. A more conservative surgical approach was devised. Indeed, the entire colon was spared and the anatomical alteration involving the valve was modified.
The successful experience with this original surgical procedure has induced us to report this new surgical approach for BVS.

Patients and methods

A 36 year old woman, married and with one son, came to our Surgical Unit on 9/7/1998 because since December 1997 she suffered from episodes of abdominal pain which was diffused mostly in the superior abdominal regions, with nausea and diarrhea, but without fever. The crises usually appeared late after meals. The occasional vomiting was not alimentary. Blood in stools has never been evidenced.
The patient's past history was free of any disease involving the abdomen and the digestive system. Abdominal barium x-rays apparently showed a true diverticulum of the transverse colon and a diverticulosis of the left colon. Antispastic treatment was ineffective. Furthermore, the patient maintained her weight and strength and was in good general conditions. At entrance in the Unit, examination revealed only slight pain after deep palpation of the right iliac fossa.
The patient underwent: ultrasounds, abdominal double contrast barium x-rays (Fig. 1 a, b - 2 a, b), CT scan without and with contrast means, and with insufflation (Fig. 3 a, b, c, d).
The x-rays evidenced a filling defect with smooth well-limited margins at the medial wall of the caecum corresponding to Bauhino's valve. This picture was considered non malignant.
At the proximal end of this zone a linear opacity similar to a chord was observed, which was due to incomplete filling of the intestinal lumen and this was interpreted as terminal iliac spasm. In the tightened zone the proximal tenuis was not dilated.
CT scan confirmed a large filling defect of the caecum at the superior tract corresponding to ileal confluence and with a very particular morphology.
No signs of cancer infiltration were reveled but a thickening of the caecum wall was seen. Blood examinations were normal.
On 9/18/1998 the patient was taken in the operating room with an explorative program.
At laparotomy ileocaecal area showed edematous tissues up to the caecum and up to the last part of the ileum, and normal serous coating. Palpation of the caecum which was slightly hyperhemic, revealed a solid mass which had soft parenchymatous consistence.
The mass was fixed to the bowel wall and difficult to interpret. At explorative caecotomy Bauhin's valve appea-

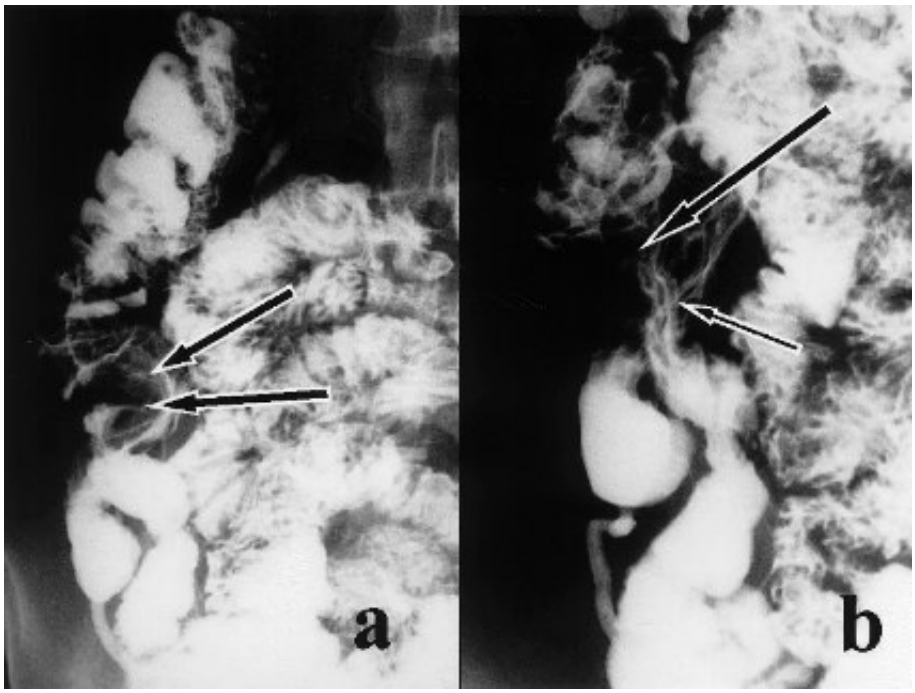


Fig. 1: a e b) Abdominal barium x-rays (8/7/1998). Terminal tract of the ileus reveals a slight mucous lining. The first tract of the ascending colon shows an apparently "extrinsic" filling defect riverberating on the terminal ileum. Caecum and appendix are full of barium. This latter aspect suggests an obstacle for contrast means to pass from caecum to ascending colon. The mucus lining lacks infiltrating dismorphism.

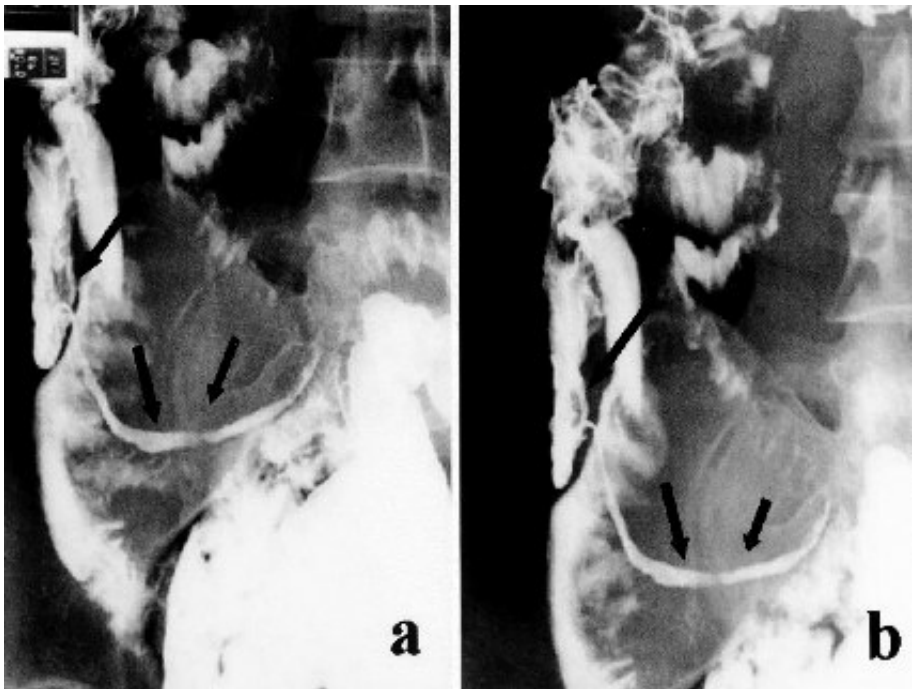


Fig. 2: a e b) Abdominal barium x-rays (8/7/1998). Prolonged spasm of the last tract. The filling defect between caecum and ascending colon is reduced.

red extremely hypertrophic and elongated (about 4 cm) soft and parenchymatous and apparently without a muscular structure (Fig. 4 b, c). A Foley catheter n° 16 (Fig. 4 b) was introduced into the ileal tract. The catheter was blown with 5 cm of H_2O. An abnormal sphincter was revealed.

The catheter was put under light traction so to block ileal transit in the caecum (Fig. 4 a).

Finally, a circular dissection of the entire hypertrophic portion was performed. This latter appeared to be made up of a double mucosal coating with an adipose content (Fig. 5 c) e d).

The two residual mucosal margins were sutured with an interrupted suture, completing with a double layer caecorrhaphy (Fig. 4 d). Suspecting that the patient's symptoms could also depend on ileocaecal chronic invagination, the last portion of the ileus was fixed on the caecum with a gun barrel modality with three seromuscular stitches. The operation was completed with appendectomy.

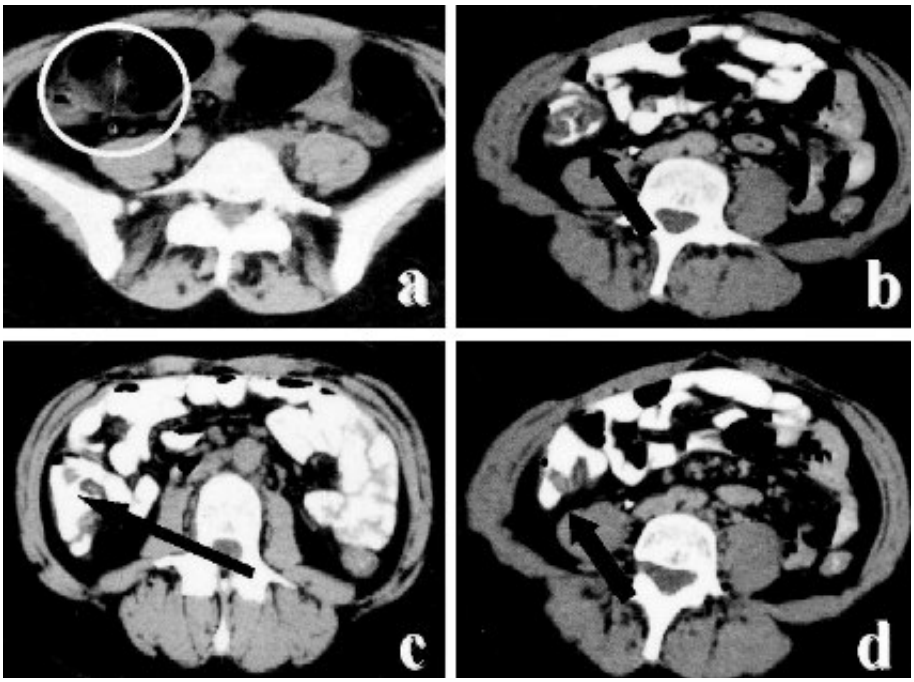


Fig. 3: a) Abdominal-pelvic CT scan without luminal contrast means. The caecum shows protrusion of a vegetating mass with clear margins. An intestinal tract on the external part of the caecum shows thick edematous walls; b) abdominal-pelvic CT scan with barium. The caecum shows an endoluminal "body" with smooth surfaces; c) Abdominal-pelvic CT scan with barium. The caecum filling defect is in turn site of barium passage; d) protrusion of the valve in caecal lumen.

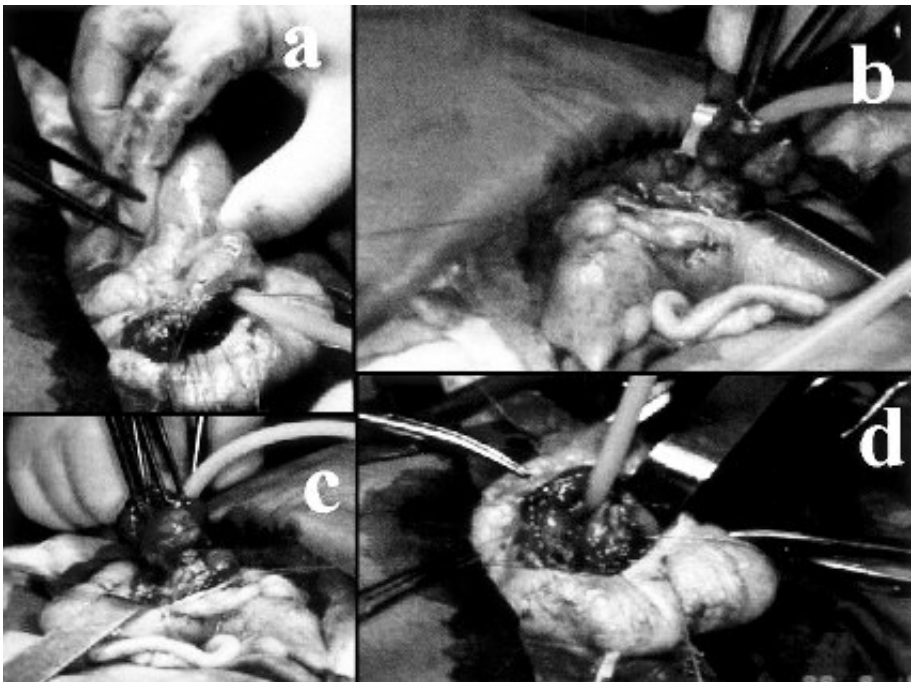


Fig. 4: a) Foley's balloon fixed before ileo-caecal sphincter; b) the catheter is put under light traction so to block ileal transit in the caecum; c) the hypertrophic structure was dissected concentrically at the base. Sectioning of hypertrophic wall has not encountered muscular structures and shows prevalence of adipose tissue in the submucosa; d) all thickness suture at the dissection base.

Results

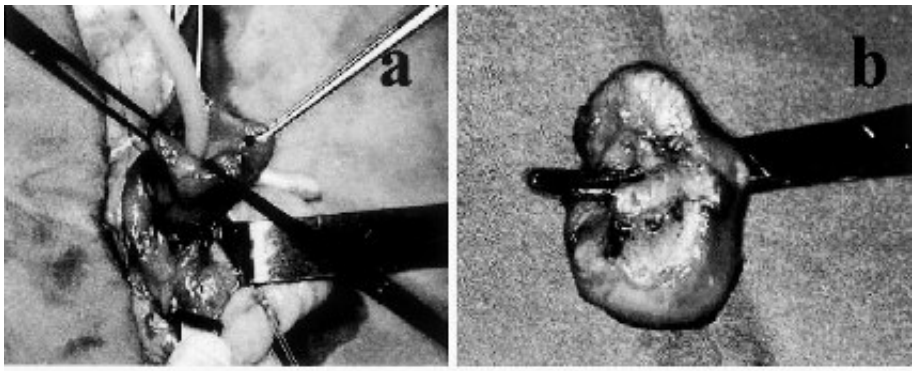
Macroscopic examination: the light yellow vegetating anular neoformation was 4 cm in its maximum diameter, showed adipose tissue extensively diffused in the submucosa, but not in the muscular layer, with clear clivage line (Fig. 5 a, b, c).

Histologic examination: multiple histologic sections showed nodosity with defined margins, made up of mature adipocytes mixed with numerous small and irregular blood vessels showing various shapes and thin walls. Adipocytes did not show cellular atypia. The post-operative period was normal, and the patient was discharged after 10 days. During this period all symptoms disappeared with full well-being of the patient.

Eight months later a barium meal follow-through was performed.

This showed a normal caecum, an ileocaecal valve incontinence with moderate reflux in the ileus (Fig. 6 a, b).

Follow-up of the patient has confirmed success of our conservative choice because of the permanent total well-being with disappearance of symptoms at forty months from operation shows that pathophysiology of BVS was obstructive and not due to reflux.



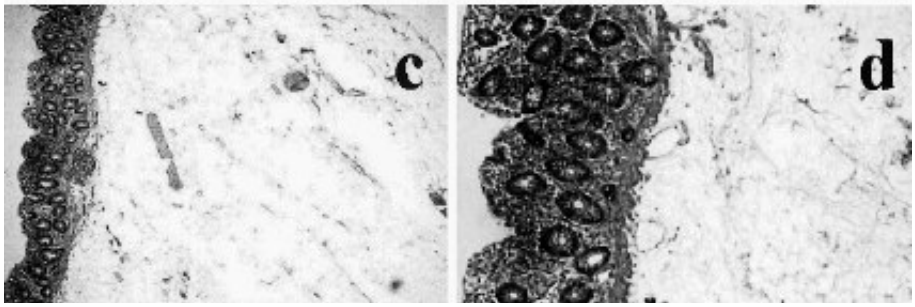


Fig. 5: a, b, c) Macroscopic and microscopic findings. Normal mucosa of the B.V. infiltrated with mononuclear cells. Thick submucosa made up of mature adipose tissue.

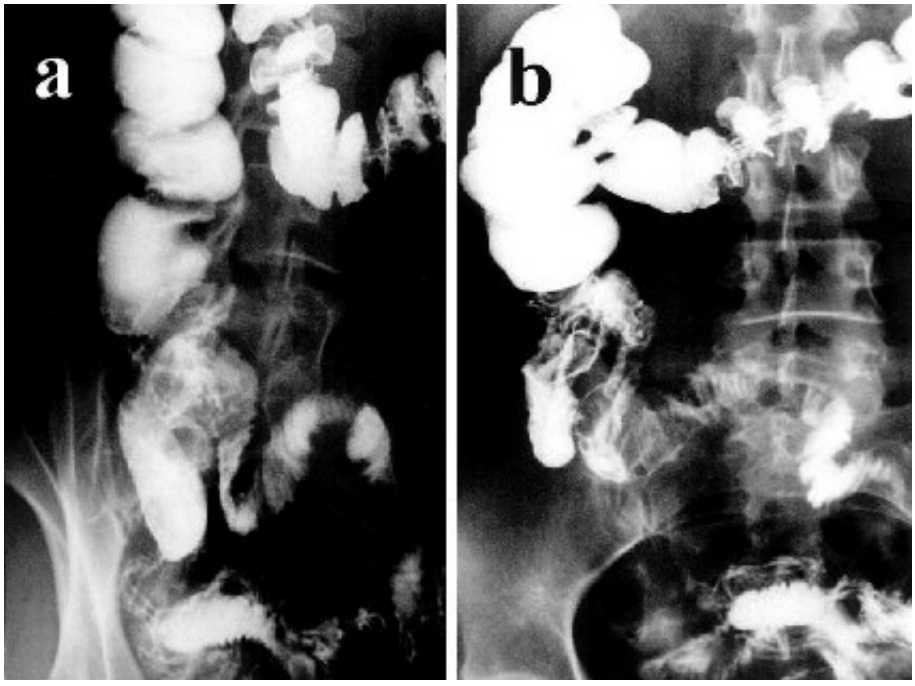


Fig. 6: a, b) Abdominal x-rays after 8 months from the operation. Large colon-ileal reflux without ileal spasms and without filling defects in the caecum-colic area.

Discussion

Literature review has evidenced controversy concerning BVS etiology. Amebiasis (Gandolfo-Canessa, 1950), regional enteritis (Strombeck 1941) and Golden 1943 were also the first to describe an x-ray picture of valve enlargement), adipose infiltration and submucosal lipoma (Edwards and Zangara, 1951) and Grayson 1958, Salem and McGee 1959), were considered responsible.(1) Lasser and Rigler (1955) (10) believed that multiple factors could be responsible for valve enlargement:

- Edema – either idiopathic or post-traumatic
- Ileal mucosa invagination into the caecum
- Fat accumulation in the submucosa
- Benign and malignant tumors
- Inflammatory lesions involving the valve.

The large number of etiological factors hypothesized suggests that the primum movens of the syndrome could be an alteration of the normal physiological activity of the ileocaecal junction. Indeed, no real valve exists and ileocaecal junction is protected only by the activity of the sphincter. Therefore, many explanations exist for this syndrome.

It is possible that the primitive lesion produces disorga-

nization of the sphincter such as incompetence or obstruction. Therefore, a non-specific inflammation of caecal mucosa around ileocaecal junction could produce an increase in sphincter activity through a nervous reflex. A sphincter spasm could produce edema, mucosa prolapse or inflammation of ileocaecal mucosa (12, 13).

Such ileocaecal sphincter hyperactivity could be associated with an increase in terminal ileum activity so to counteract sphincter spasm (14, 15). This could justify pain and diarrhea.

The fact that diseases such as malignant and benign tumors (16, 17) have been evidenced in patients suffering from BVS suggests that the alterations produce sphincter spasm which in turn triggers the events observed in this disease.

Therefore, the final hypothesis is that BVS is due to sphincter hyperactivity which is often secondary to mucose lesion around ileocaecal junction (18, 19).

In our case lipomatous hypertrophy and abnormal elongation of ileus with ileal sphincter in situ in the caecum and ileal spasm corresponding to clinical localized abdominal pain and pain after palpation of right abdominal regions. Such crises were associated with typical symptoms of sympathetic hypertone such as sweating and nausea, and sometimes non alimentary vomiting i.e. away from meals.

Crises were followed by diarrhea with semiliquid stools which suggest a rapid emptying of the tenuis in the caecum and therefore colon hyperperistalsis.

The entity of the caecum filling defect often detected on preoperative x-rays varied greatly in time and a partial ileocaecal intussusception with an apparently normal ileal sphincter (as seen during introperative transcaecotomy) could not be excluded.

Furthermore, more than one anatomical-clinical condition exists underlying BVS which is characterized by a periodic obstacle to ileo-caecal transit.

This represents the pathophysiological basis for pain and diarrhea. Since BVS is a disease localized in the submucosa of an hypertrophic valve, then right hemicholectomy, constantly described in literature, does not seem indicated since it appears too destroying for a non-malignant disease.

Conclusions

Reports dealing with BVS are scarce in literature and our single experience is naturally not enough to trace absolute guidelines.

In view of the good results obtained by means of this original operation, three key aspects of the procedure seem noteworthy:

- Endocaecal exploration for caecotomy, after adequate antibiotic and mechanical preparation of the colon;
- Hypertrophic valve resection downward with respect to the ileocaecal sphincter evidenced by on the balloon of Foley's catheter located in the ileus passing from the caecum;
- Seromuscular fixation of the last part of the ileum to the caecum in order to prevent chronic intussusception.

Whether unsuccessful, this surgical procedure could still be followed by successive hemicholectomy or ileocaecal resection, necessary in this case.

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