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Lane's Disease – Delayed Transit Idiopathic Constipation

Summary

Lane's disease occurs only in young women and is associated with severe constipation and various diseases such as peripheral circulatory disorders, orthostatic hypotension, water retention, urinary difficulties and gynaecological disorders. The last include amenorrhea, galactorrhea, infertility, loss of secondary sexual characteristics and polycystic ovary. Constipation is always present in this syndrome and stems from delayed transit of faeces through the large bowel together with possible disorders of defaecation mechanism. The diagnosis, aetiology and medical and surgical treatment are described.
(Keywords: Lane's disease, constipation)

Résumé

Le Morbus Lane est une maladie qui apparaît exclusivement chez les femmes jeunes. Elle se traduit par une constipation sévère, des perturbations de la circulation périphérique, une hypotension orthostatique, une rétention d'urine, des troubles de la miction et des problèmes gynécologiques: aménorrhée, galactorrhée, stérilité, perte des caractères sexuels secondaires, polykystose ovarienne. Dans tous les cas la constipation est présente, elle résulte d'un transit des selles retardé au niveau du gros intestin et d'éventuels troubles du mécanisme de la défécation. Les auteurs décrivent le diagnostic, l'étiologie et les traitements médicaux et chirurgicaux de cette maladie.
(Mots clés: Morbus Lane, constipation)

Resumen

La enfermedad de Lane se presenta sólo en mujeres jóvenes y va unida a un fuerte esteñimiento y varias dolencias más, como trastornos circulatorios periféricos, hipotensión ortostática, retención de agua, dificultades urinarias y trastornos ginecológicos. Estos últimos comprenden amenorrea, galactorrea, esterilidad, pérdida de características sexuales secundarias y ovario poliquístico. En este síndrome siempre hay estreñimiento y en el intestino grueso detenciones del tránsito retardado de las heces junto a posibles trastornes en el mecanismo de la defecación. Se describen el diagnóstico, la etiología y el tratamiento médico y quirúrgico.
(Palabras clave: Enfermedad de Lane, estreñimiento)

Introduction

Today functional, chronic constipation in the adult tends still to be superficially regarded and therefore the patient's complaints may be underestimated, treatment incongruous and may be abandoned after a long, difficult relationship with the doctor. Constipation should always be considered important not only because it is widespread in western countries, but also because of the psychological problems the more severe form can cause. The pathogenesis of the disease is complex and requires a careful individual study in order to assess the particular aspects that are needed for correct treatment. The occurrence of unusual functional constipation in association with other symptoms constitutes a rare and little known syndrome – the Arbuthnot Lane's disease.

As early as 1909 Lane [1] published a paper titled »Chronic intestinal stasis« in which he pointed out the frequent coexisting of intractable severe constipation

with genital disorders such as infertility, amenorrhea, increased incidence of ovarian cysts, breast nodules and disorders of other systems such as peripheral circulation, excessive skin pigmentation, and urinary difficulties in young women. However, his explanation that the disorders were associated with toxin absorption from constipated colon and consequent advocacy of total colectomy were less acceptable. However, both these studies and the name attributed to them soon became obsolete. Recently, after the introduction in 1969 of Hinton's method of studying intestinal transit time, Lane's syndrome was rediscovered, and numerous series have been reported since 1980. These include a series of 54 cases by Waiter et al. [2]; 25 cases by Lanfranchi et al. [3]; 75 cases by Preston and Lennard-Jones [4]; 26 cases by Schuffler et al. [5]; 14 cases by Read et al. [6] and 25 cases by Shouler and Keighley [7]. These studies have set down a rational, scientific basis for the syndrome even though many of its aspects are still not clear. We report two cases in young women with severe constipation whose diagnosis of Lane's disease was achieved by the most up-to-date methods and techniques used.

Case Reports

Two young women, aged 20 and 18 years respectively, came to our department complaining of severe constipation that could only be partly alleviated by massive administration of laxatives. They could only perform one, or infrequently, two evacuations each week after large doses of intestinal smooth muscle stimulating drugs. Without drugs this interval reached two weeks or more. Moreover they reported having abdominal distension, nausea, anorexia and felt unwell. The onset of constipation was at puberty with progressive worsening and reaching a peak after surgery for umbilical hernia in one case and for appendicitis in the other. They admitted that the constipation had a psychological effect on their lives. Other disturbances were also present in both patients such as difficulty in micturition which sometimes resulted in total blockage, prolonged amenorrhea and circulatory deficiency to the extremities attributed to Raynaud's syndrome.
An X-ray study of the colorectum was carried out with barium enema and revealed regular shape and normal size of the colon and rectal ampulla (Fig.1). Therefore localised lesions causing strictures and functional diseases such as Hirschsprung's disease were ruled out. Anorectal manometry (Fig.2) which revealed completely normal anorectal inhibition reflex and histological examination of an endoscopic biopsy furthermore ruled out congenital aganglionosis of the colon. This

Fig.1: Barium enema: regular shape and normal size of the colon and rectal ampulla

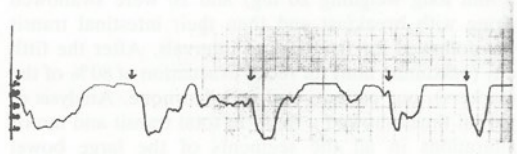

Fig.2: Anorectal manometry: normal anorectal inhibition reflex

biopsy confirmed the presence of quantitatively and morphologically normal gangliar cells. Transit times were then studied according to Hinton's method. The radio-opaque markers conformed to the standard required (segments of radio-opaque polyethylene tubes

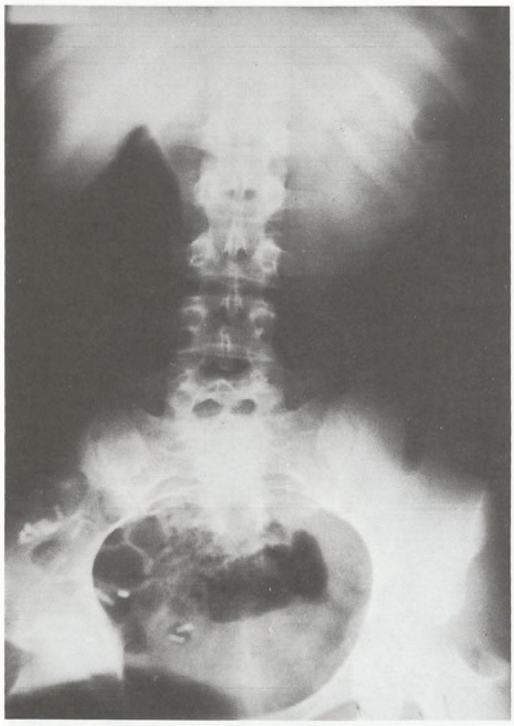

Fig. 3: Transit times of the colon. Markers at 6th hours

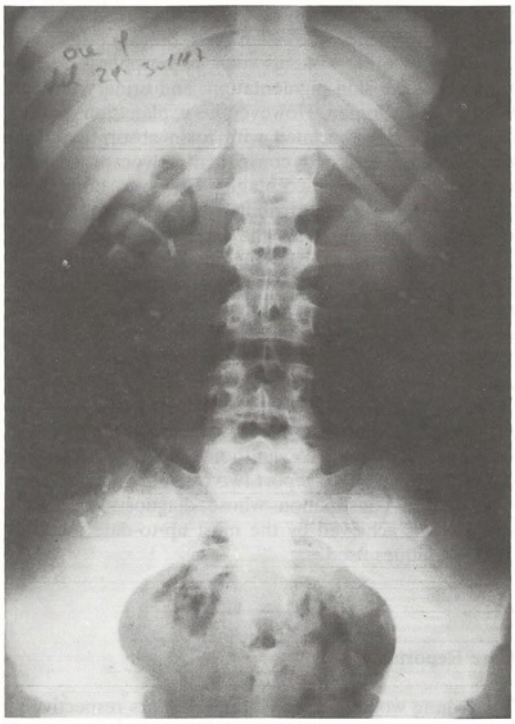

Fig. 4: Transit times. Markers at 48th hours

3 mm long weighing 20 mg) and 20 were swallowed along with breakfast and then their intestinal transit was followed for five days at intervals. After the fifth day (maximum limit for rectal evacuation of 80% of the markers), our patients had expelled none. Analysis of partial times showed a delay in total transit and motor alterations in all the segments of the large bowel (Figs.3,4,5). Study of the motility index (MI = n wave's amplitude x duration/time) obtained with the use of a 70 cm polyethylene probe with three recording points allowed us to quantify the motor deficit (< 40% in the first and < 30% in the second patient). Functional tests revealed in one patient abnormal defaecation which often accompanies delayed intestinal transit, evacuation being well over the normal five minute limit. These examinations revealed deficient sensation (first sensation at 140 ml of a distending ballon) and a failure to relax the striated muscles of the pelvic floor and external anal sphincter resulting in inability to expell a balloon containing 50 ml of water (easy for a normal patient) from the rectum. Electromyographic examination of the external sphincter confirmed paradoxical contraction of the striated muscles at defaecation (Fig.6). A form of »slow transit disease« was diagnosed in each and further examinations were carried out. Study of the genital system showed hypogonadotrophic hypogonadism and micropolycystic ovary. Hyperprolactinaemia was also found in one case.

Discussion

The severe constipation in these patients is caused by the delayed transit of faeces through the bowel. This is demonstrated by the marked delay in transit of the markers through the bowel while passage through the small intestine is within the normal time limit. The inertia of the bowel is widespread and not limited to specific sectors. In numerous cases constipation is aggravated even further by the presence of the so-called

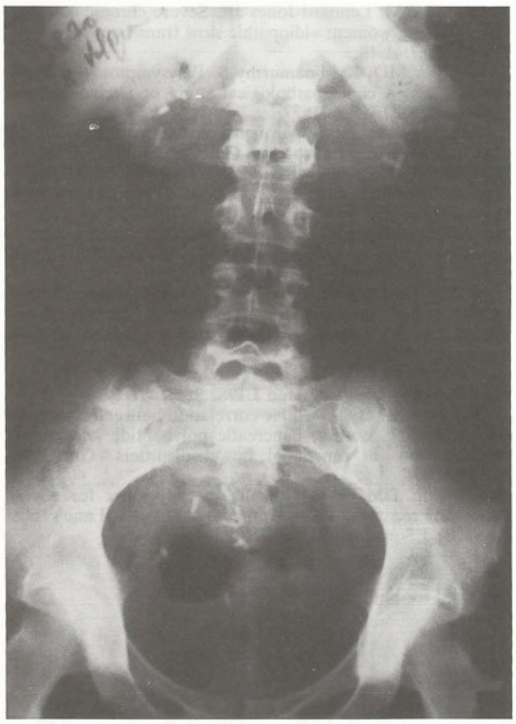

Fig. 5: Transit times. Markers at fifth day

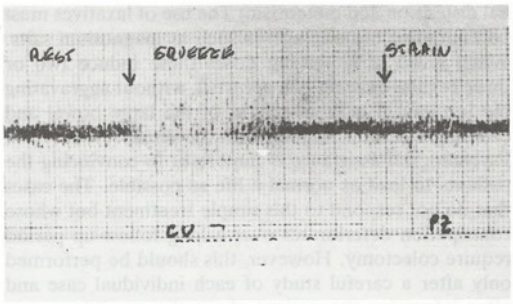

Fig. 6: EMG of external sphincter: paradoxical contraction on straining

»rectal blockage« which causes prolonged straining during defaecation. This can be caused by many factors including various mechanisms such as increased anorectal inhibition reflex threshold, augmented stimulus sensitivity (rectal dischezia) and above all paradoxical contraction of the puborectal muscle (anism). If these are the main characteristics of this particular type of constipation, many questions remain unanswered. What is the cause of the bowel inertia? Is there a connection between it and the rectal blockage or is this association just a coincidence? Is there a common denominator which can explain the association with the other disorders, especially with the gynaecological disorders?

At present no definite answers can be given, nevertheless some rational hypotheses can be put forward. Atrophy of the myoenteric plexus in the colon is often present in patients whose severity of the constipation called for surgery and some authors [8] believe this is pathognomonic of the disease. However, it is difficult to distinguish it from the lesions caused by laxative abuse.

There is also no possible interpretation for the association of colon inertia and rectal blockage. The presence of a common denominator can be deduced from the widespread motor disorders which can involve the muscles of the pelvic floor, the oesophagus and the bladder as well as the colon. The fact that Lane's disease is found only in women and the coexistence of genital diseases (infertility, polycystic ovary, breast nodules, galactorrhea etc) could indicate a hormonal common denominator of the disorders. It seems that the altered sexual hormone secretion influences the intestinal motility. Lawson et al. [9] attributed the lengthened colon transit time during pregnancy to the elevated progesterone levels which, according to Christofides et al. [10] could have an inhibitory action on secretion of motilin. Preston et al. [11] noted low levels of motilin, gastrin and pancreatic polypeptides in patients with colon inertia. Another hypothesis suggests that the motor disorders and the Raynaud's syndrome could be caused by alteration of the autonomic system.

The aetiopathogenesis of Lane's disease is controversial, therefore attention must be focused on the best type of treatment to alleviate the patient's distress. Since it is known that the altered colon motility is irreversible, treatment must be aimed at stimulating regular emptying of the bowel by means of suitable laxatives which have no serious side effects. We must avoid massive daily doses of intestinal smooth muscle stimulants (anthraquinones, sennosides, bisacodyl) which provoke a deterioration of the nervous plexus of the intestinal wall with inevitable worsening of the constipation and opening of a vicious circle that leads to ever increasing doses of laxatives. Ingestion of bran and fibre should be avoided as they do not help constipation, but on the contrary cause nausea due to abdomi-

nal distension and meteorism. The use of laxatives must be limited to osmotic agents such as magnesium salts. When given in increasing doses these induce two or three regular evacuations per week without aggravating the intramural nervous plexus of the large bowel and ensuring a certain stability with the disease.

Psychological assistance is important in convincing the patients to lead as normal a life as possible. The cases that do not respond to this simple treatment but whose constipation deteriorates over a long follow-up period require colectomy. However, this should be performed only after a careful study of each individual case and after informing the patient about the possible consequences. Since the whole colon is involved, total colectomy is required with caecal- or ileorectal anastomosis. Attempts at partial resection do not give satisfactory results. Preston's [12] limited series reports satisfactory results in the majority of the cases, in fact many unhappy women were able to revert to almost normal conditions. However, a percentage of women remained with persistent constipation and others with excessive daily evacuation. The fact that in certain cases the only solution to severe constipation is radical surgery despite the youth of the patient, the absence of organic pathology and the controversial results obtained underline the necessity of paying greater attention to the problem of constipation.

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