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Digestive Diffuse Angiomatosis, an Exceptional Cause of Intestinal Occlusion: About A Case

I.Bouhout, T.Bouhout, R.Jabi and M.Bouziane

Department of general surgery, Mohammed VI University Hospital, Faculty of Medicine and Pharmacy of Oujda, Mohammed First University, Oujda. Morocco.

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ABSTRACT

Blue rubber bleb nevus syndrome (Cutaneous and digestive angiomatosis) is a vascular malformation that is not well known. The gastrointestinal lesions occurring during this syndrome are clinically more relevant than those of the skin and soft tissues. Adherence to standard surgical principles results in successful results with minimal morbidity. The diagnosis of Bean syndrome must be evoked in the presence of acute intestinal occlusion and angiomatous skin lesions. We report a particular case of this syndrome by its late revelation following a volvulus on a huge bowel vascular lesion, giving the opportunity to a review of the literature.

1. Introduction

Bean syndrome (Cutaneous and digestive angiomatosis) is a vascular malformation that is not well known. It is revealed especially from the first years of life. It is manifested by angiomatous cutaneous and visceral lesions that are frequently associated with bleeding and anemia that can be fatal. [1]

Typically, skin lesions are usually asymptomatic, easily compressive. For clinicians, gastrointestinal bleeding and secondary iron deficiency anemia are the most common symptoms of gastrointestinal tract damage. [2]

We report a particular case of this syndrome by its late revelation following a volvulus on a huge glandular vascular lesion, giving the opportunity to review the literature.

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2. Observation

The case of a 70 - year - old patient, known to have multiple cutaneous angiomas at an early age, is admitted to the emergency room in an acute intestinal obstruction chart.

On clinical examination, the patient was hemodynamically, respiratory and neurologically stable with diffuse abdominal distension and multiple bluish angiomatous lesions easily compressible on the palpation distributed diffusely and symmetrically on her body without any other features under examination. clinical. (Figure 1)

An unprepared abdomen was made and objected to hydro-aeric levels of the grelic type. The abdominopelvic CT showed a dilated flat / dilated small junction with a Whirl Sign appearance suggestive of hail volvulus (Figure 2)

An exploratory laparotomy demonstrated small bowel volvulus on diffuse angiomatosis spread over two and a half meters of hail (Figure 3). No resection was necessary. The postoperative course was simple with full recovery of the patient.

The diagnosis of Bean Syndrome was found in association with diffuse angiomatous lesions and hail volvulus. A complementary assessment of Bean Syndrome associated lesions was performed and returned with no particularities.

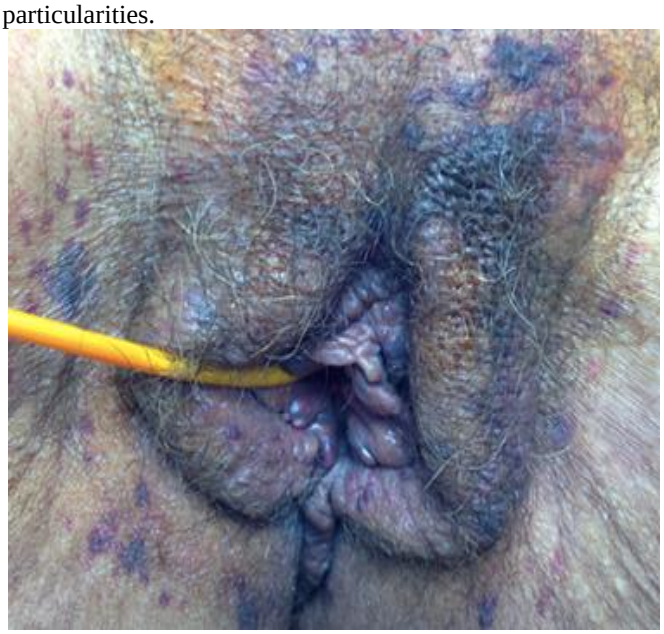

Figure 1. Image showing multiple bluish angiomatous lesions on the anogenital region of our patient.

Tele: 00212672854691

E-mail address: dr.imad.bouhout@gmail.com

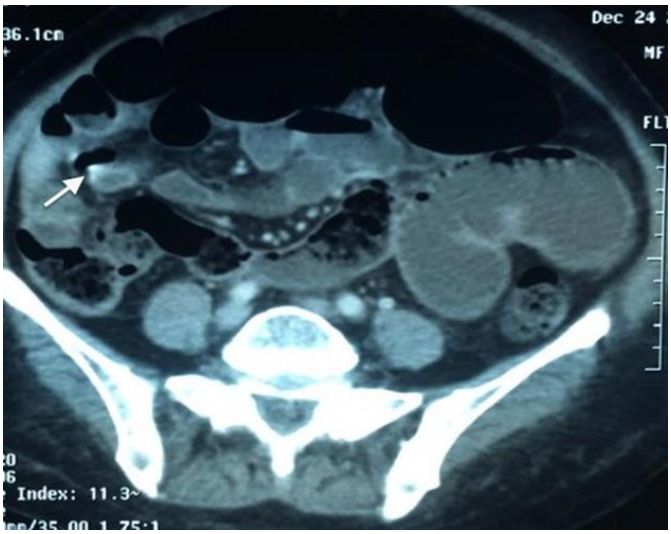

Figure 2. Scanning image showing contrast enhancement by a vascular lesion at the level of the pelvic wall in relation to pelvic angioma.

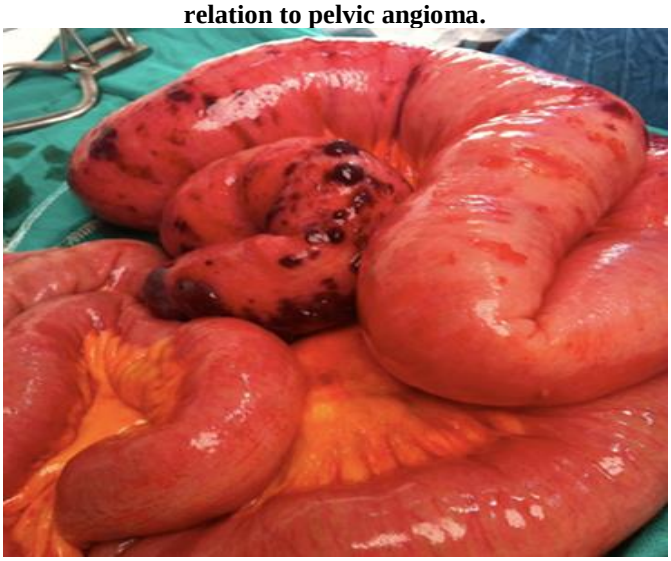

Figure3. Intraoperative image showing multiple angiomatous lesions of the pelvic wall.

3. Discussion

Although William Bennett Bean drew attention to the syndrome that bears his name in 1958, Gascoyen described it almost 100 years ago [3].

The cause of Bean syndrome is still unknown. However, the locus on chromosome 9p and the high expression of c-kit can be involved at the molecular and genetic level. [4]

The majority of cases appear to be sporadic although autosomal dominant transmission has been described in several familial cases, [5]

Venous malformations are multifocal and most often affect the skin and the digestive tract. In a literature review of Xue-Li Jin about 120 cases, cutaneous angiomas were observed in 93% of cases, and gastrointestinal lesions in 76% of cases [6] with a positive correlation between the number of cases. gastrointestinal lesions and the number of cutaneous haemangiomas. [7]

Other parts of the body may have been affected but less frequently such as liver, spleen, pancreas, kidneys, bladder or eyes. [8,9]

Digestive hemorrhage is the most common mode of disclosure.

Usually, gastrointestinal lesions occur at an older age with intermittent melena and massive rectorrhagia being rare manifestations. Exceptionally, abdominal pain, intussusception, volvulus, infarction can occur. [10,11]

Our patient had presented a volvulus as a way of revealing Bean syndrome, making it a unique case.

Venous malformations can occur interchangeably along the gastrointestinal tract, from the oral mucosa to the anal canal, and are primarily of interest in the small intestine and distal colon. Some studies have reported that the small intestine is the most involved extracutaneous localization. [12]

Biologically, iron deficient hypochromic microcytic anemia, coagulopathy and / or positive blood culture can be observed. [2].

Computed tomography, particularly with contrast injection, is the most invaluable non-invasive diagnostic tool for BEAN syndrome, which identifies gastrointestinal hemangiomas and extra-intestinal lesions. [2]

In case of absence of cutaneous lesions, the diagnosis is usually made only intraoperatively.

Complications like volvulus or intussusception are exceptionally revealing. In these cases, when the lesion is localized, resection of the small bowel is recommended. In our patient, it was an extensive lesion with major distention of the first jejunal loop. An extensive resection of the hail seemed inappropriate. [13-15]

In the absence of significant intestinal bleeding, corticosteroids, antifibrinolytic agents, intravenous immunoglobulin in high doses and interferon alpha are not very effective. [13-15]

4. Conclusion

Gastrointestinal lesions in Bean syndrome are clinically more relevant than skin and soft tissue lesions. Although surgery can be demanding and tedious, a good multidisciplinary approach with adherence to standard surgical principles leads to successful outcomes with minimal morbidity.

The diagnosis of Bean syndrome must be made in the presence of acute intestinal obstruction and angiomatous skin lesions.

Conflict of Interest

The authors do not declare any conflict of interest.

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