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Bean Syndrom: About an Observation and Review of the Literature

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Abstract – We report the case of a 14-year-old girl who presented chronic anemia after a digestive hemorrhage episode. Clinically, she presented other sites manifestly of vascular proliferation (vulva). Upper digestive fibroscopy showed a very hemorrhagic polyp of the greater curvature of the stomach, and another of the duodenum. The jejunal tumor was removed for pathological examination. The specimen was a pedunculated polypoid fragment measuring 1.5 cm of long axis. The section showed polycystic section with brown contents. The histological diagnosis was a cavernous hemangioma.
Taking into account clinical, endoscopic, laparoscopic, and histopathological datas, the diagnosis of Bean syndrom was retained.

Keywords – Bean Syndrom, Madagascar, Cavernous Hemangioma, Jejunum, Pathology

I. INTRODUCTION

Bean syndrom, also known as "Blue Rubber Bleb Nevus Syndrome" (BRBNS) is a disease characterized by multiple vascular malformations that affect the skin, the gastrointestinal tract and less often other viscera [1]. It is a rare condition. Actually, more than 200 cases have been described, but the prevalence remains unknown [2, 3].The aim of this study is to report a Malagasy case and compare it with cases from the literature.

II. OBSERVATION

It was a 14-year-old girl who presented chronic anemia after a digestive hemorrhage episode. Physical examination showed in the hypogastric region a surgical scar from an anterior vascular tumor, and a vascular spot on the vulva. Upper digestive fibroscopy showed a very hemorrhagic polyp of the greater curvature of the stomach, and another of the duodenum. The laparotomy showed a vascular tumor of the liver, and a very hemorrhagic jejunal tumor. Only the jejunal tumor was removed for pathological examination.

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The specimen was a pedunculated polypoid fragment measuring 1.5 cm of long axis. The section showed polycystic section with brown contents. The histological examination objectified an intestinal mucosa with villi and glands lined with cells devoid of cytonuclear atypia. The submucosa presented a venous vascular proliferation, lined with non-atypical endothelial cells. The vascular lumens were dilated and intercommunicating, containing red blood cells. The histological diagnosis was a cavernous hemangioma, part of the slow flow vascular malformations.

Taking into account clinical, endoscopic, laparoscopic, and histopathological datas, the diagnosis retained was Bean syndrom, also called "Blue Rubber Bleb Nevus Syndrom".

After the diagnosis, the genetic investigation mentioned the absence of a similar case in the family.

III. Discussion

The Blue Rubber Bleb Nevus (BRBNS) syndrom was described by Gascoyen [4] a century before William Bennett Bean in turn reported it and named it in 1958.

It is a rare congenital condition, but its incidence is still little known. In the literature, the authors only report clinical cases. To date, 200 cases have been reported [2, 3]. To our knowledge, no Malagasy case has yet been reported.

Bean syndrom is characterized by a disseminated and sporadic venous malformation but could be familial with autosomal dominant transmission secondary to a mutation on chromosome 9p [5]. The pathogenesis is still less known.

Our case presented a cutaneous, digestive, and hepatic attack. In the literature review, the cutaneous and digestive localizations are the most observed [6].

The skin localization is the most frequent. The study by Xue-Li Jin [8] on 120 cases of literature review mentions that the skin is affected in 93% of the cases. It is observed during the physical examination and can affect the whole body but often the trunk as in our case, and the upper limbs. The skin lesions are multiple non-bleeding and can be macules, papules, or nodules, of reddish or purplish color. They are present at birth or appear over the years [7].

Digestive localization is also common. Xue-Li Jin [8] mentions a percentage of 76% of the cases. It is clinically manifested by digestive haemorrhages which are the main complications of this disease (occult bleeding, hematemesis, melena, or rectorrhagia). These digestive haemorrhages encourage patients to consult. Endoscopy with biopsy is the first line for diagnosis. The lesion can be polylobulated, nodular, sessile, pediculated, or ulcerated [9]. The mucous membrane of the digestive tract, from the mouth to the anus is concerned, but the small intestine is the site most frequently reported [9], as in our case, and those of the literature.

Other localizations are reported: the liver as in our case, the spleen, the pancreas, the kidney, the bladder, the eye, the brain, the heart, the lung, the muscles, the peritoneal cavity, the thyroid, the thymus, submandibular and parotid regions, and joints [6]. Depending on the location, the lesions may be responsible for orthopedic deformation, dementia, pulmonary embolism, hypertension of the pulmonary artery, hemothorax, hemopericardium, exophthalmos [8, 10, 11, 12].

From an anatomopathological point of view, the lesion is composed of a vascular proliferation, of various sizes, sometimes cystic and inter-communicating. The lumen contains red blood cells, and is lined with non-atypical endothelial cells. The wall is made up of pericytes, smooth muscle fibers, and fibroblasts [13]. The malignant transformation of lesions has so far not been described [2].

In the absence of appropriate management, affected patients suffer from anemia due to chronic bleeding which can be fatal.

Nowadays, there is no curative treatment for Bean syndrom. Iron administration and repeated transfusions correct anemia if there is no significant bleeding. According to Achraf El [2], surgery with excision of intestinal angiomas by laparoscopic or conventional route, seems to be the effective means to improve the prognosis of children with this syndrom and thus reduce the frequency of transfusions. Zhi Li [14] also admitted that surgical eradication of intestinal angiomas is an effective way to control severe chronic intestinal bleeding regardless of its number or location and this by resection of all lesions responsible for bleeding as well as segments digestive sites of lesions of doubtful viability. In our case, only the bleeding intestinal angioma was removed. This allowed hemodynamic stabilization of the child without guaranteeing a relapse during bleeding from other angiomas (gastric and duodenal). Since 2012, authors have reported the effectiveness of the use of Sirolimus which is an antiangiogenic and antineoplastic immunosuppressant [15, 16]. Its mechanism of action is the inhibition of rapamycin (mTOR), which is a serine-

threonine kinase, regulated by phosphoinositide-3-kinase (PI3K) [16]. It has been used successfully in the management of several vascular abnormalities such as kaposiform hemangioendothelioma, angioma, and lymphatic malformations [16].

IV. Conclusion

Bean syndrom is a rare vascular malformation disease that should be considered. The most frequent localizations are cutaneous and digestive. The main complication is digestive hemorrhage. Other drug treatments are still being evaluated. Apart from transfusion and hemostasis surgery which provide comfort for the patient, there is no curative treatment yet.

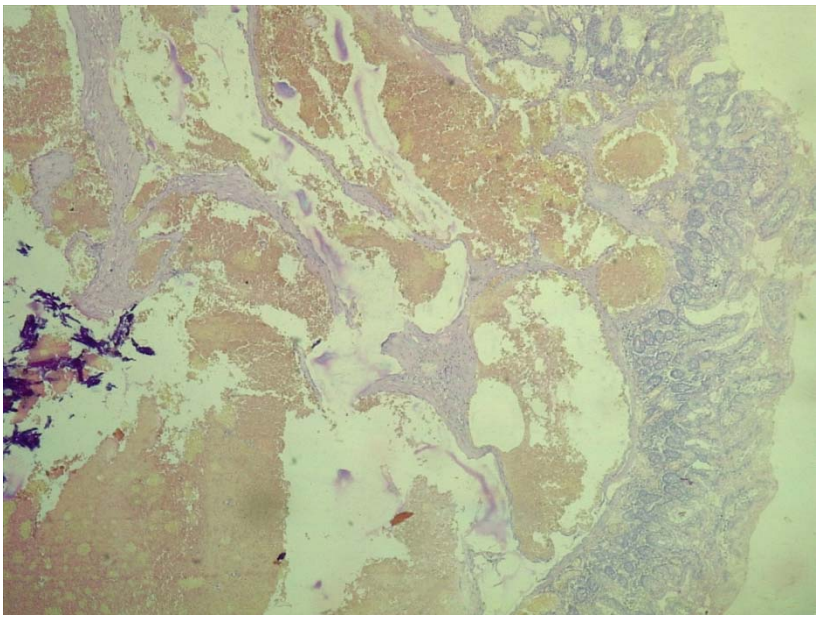

Figure 1: histological pattern of blood vessels proliferation of the ileum. HE x 4

Source: Anatomy and Pathological Cytology Laboratory of Joseph Ravoahangy Andrianavalona University Hospital

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