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GASTRIC AMYLOIDOSIS REVEALING THE DISEASE

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ABSTRACT

Amyloidosis is a heterogeneous group of diseases characterized by amyloid deposition in different organs. Digestive involvement is often asymptomatic, digestive haemorrhage is rare but can prove fatal. We report a case of a 61-year-old woman with systemic amyloidosis revealed by a multirecurrent digestive hemorrhage. Through this case and a review of the literature, we emphasize the gravity of this location and the difficulty of taking care of it

Keywords : Amyloidosis, digestive hemorrhage.

INTRODUCTION

Amyloses are characterized by the extracellular deposition of protein material in insoluble fibrillar form. The digestive impairment of amyloidosis is classic but the digestive haemorrhage is erratic.

We report here the observation of a systemic amyloidosis revealed by a digestive haemorrhage, the gravity and the difficulty of management can characterize this localization.

MATERIALS AND METHOD

Patient of 61 years, with a history of MGUS of type IgG Kappa, hospitalized for a renal insufficiency with proteinuria. During the course of her hospitalization, the patient presents a digestive haemorrhage of type melena and derectorragies.

A first FOGD showed a small angioma of the bulb without haemorrhagic sign and rectosigmoidoscopy concluded to grade 2 internal hemorrhoids.



Figure 1: angioma at the 1st FOGD



Figure 2: Hemorrhoids internal to the rectosigmoidoscopi

The examination revealed an alteration of the general state. Clinical examination was normal. Hemoglobin was 7 g / dl after transfusions.

Evolution was marked by the spontaneous cessation of bleeding, 3 days after bleeding, colonoscopy revealed a polyp of the coecum, the enteroscopy of the small bowel by videocapsule was carried out without ever identifying the endoscopic zone responsible for the bleeding, hemorrhage.

The persistence of the dislobulization imposed several series of transfusions. Medical treatment with sandostatin was proposed empirically.

A second FOGD, due to persistent deglobulation despite transfusions, demonstrating an angiodysplasia of the fundus jenus near a fibrinous ulcer forest III requiring an electrocoagulation session.



Figure 3: fibrinous ulcer at the 2nd FOGD

The anatomopathological examination of the specimen carried out showed amyloid deposits taking the Congo red at the level of the vascular walls of the submucosal vessels and a yellow and green birefringence in polarized light. Immunofixation showed AL amyloidosis.

The diagnosis of systemic amyloidosis (digestive and renal) was made.

Actiological treatment of AL amyloidosis was initiated and there was no bleeding recurrence at one year.

RESULT AND DISCUSSION

Amyloidosis is a rare but serious disease characterized by the extracellular deposition of protein material in insoluble fibrillar form. Six types of amylose are distinguished, depending on the biochemical characteristics of the protein precursors: primary, secondary, associated with hemodialysis, hereditary, senile and localized. AL amyloidosis is one of the most common. It results from the deposition of light chains of monoclonal immunoglobulins secreted by a plasma cell clone. The proliferation is either major, obvious and malignant, as is most often the case in myeloma, or rarely another malignant disease of B lymphocytes, or more difficult to detect and benign: in this case, it Is a primary or isolated AL amyloidosis. The term "primitive" is ambiguous and should be abandoned. It would seem simpler to call AL amyloidosis without further clarification of the disease when there is no association with a myeloma and in the reverse case to use the term amylose AL associated with myeloma. AA amyloidosis (or secondary amyloidosis) is associated with a normal protein present in excess due to inflammation, chronic infection or, more rarely, neoplasia. Amylose deposits are sometimes located in a single organ. In some cases, when the aspect is pseudotumoral,

the term localized amyloidosis or amyloidoma [1] is used. This necessitates the systematic exclusion of other possible localizations by an exhaustive assessment according to the organ affected. Most of these amyloids are of type AL and are the result of local and non-medullary plasma proliferation, the treatment is local (generally surgical) with cure, but some varieties of these localized forms are of type AA.

Gastrointestinal amyloidosis is very common in patients with systemic amyloidosis, but clinical manifestations affect only 30-60% of patients [2]. Gastrointestinal manifestations are considered to be less frequent in cases of AL amyloidosis, but symptomatic gastric disorders appear to be more frequent in AA amyloidosis [2].

All segments of the digestive tract can be affected. Depending on the site of amyloid deposits, the manifestations are either hemorrhagic and ischemic (vascular deposits) or motor (deposits in the nervous system) or mixed (disseminated deposits). It is therefore not uncommon for digestive haemorrhage to occur during amyloidosis due to amyloid vascular deposits, sometimes even in the absence of systemic amyloidosis [3].

Gastric involvement in amyloidosis was estimated to be 1% in a large series of patients with systemic amyloidosis [4]. In the series of 769 Mayo Clinic patients with AL amyloidosis, 8% had digestive tract lesions documented by biopsy, but only 1% were symptomatically gastric [5]. Digestive hemorrhage may be recurrent, as in our case, even massive and fatal [6-8]. Amouri et al (9) and Alneaimi et al (10) also described a digestive hemorrhage revealing the disease, as in our case. Sometimes, gastric involvement occurs in a pseudotumoral form [11]. It can sometimes impose an emergency hemostasis surgery [7,8] which is not always able to avoid death [7]. Beyazit et al. [12] reported a case where local application of a haemostatic powder in a 77-year-old patient with amyloidosis complicated by digestive hemorrhage by gastric ulceration was effective.

CONCLUSION

Digestive manifestations of amyloidosis are rare but may be severe. The symptomatology is diverse and varied, requiring a series of exploration in order to obtain an adequate management.

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