GREEN APPLE AND RED CONGO: ATYPICAL DYSPHAGIA COLOURS.

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Abstract

Amyloidosis is a rare disease that consists of the extracellular deposition of insoluble protein subunits known as fibrils, resistant to proteolytic degradation. It can occur in any organ of the body, causing its slow and gradual failure.

Specific involvement of the gastrointestinal tract interferes with its structure and function, most commonly in the liver and small intestine, with esophageal involvement being a rare entity, and whose main clinical manifestation is reflux. On the other hand, presentation in the form of dysphagia is highly uncommon.

Keywords: esophageal amyloidosis, dysphagia, congo red.

Clinical Case

71-year-old male, hypertensive and dyslipidaemic. At the age of 40, he was referred to the otorhinolaryngology department for his debut with dyspnoea, and was diagnosed with laryngeal amyloidosis. At 70 years of age, he began with progressive dysphagia to solids and liquids, denying pyrosis, weight loss, abdominal discomfort or other symptoms, with unremarkable blood tests. Esophagogastroduodenoscopy was performed, showing granular esophageal mucosa (Figure 1), from which biopsies were taken. Barium swallow was also performed, which showed no evidence of swallowing disorder. The biopsies showed submucosal deposits of amorphous eosinophils, positive for Congo red staining (Figure 2), and serum amyloid P, with green birefringence by polarised light microscopy (Figure 3), these findings being compatible with the diagnosis of esophageal amyloidosis.

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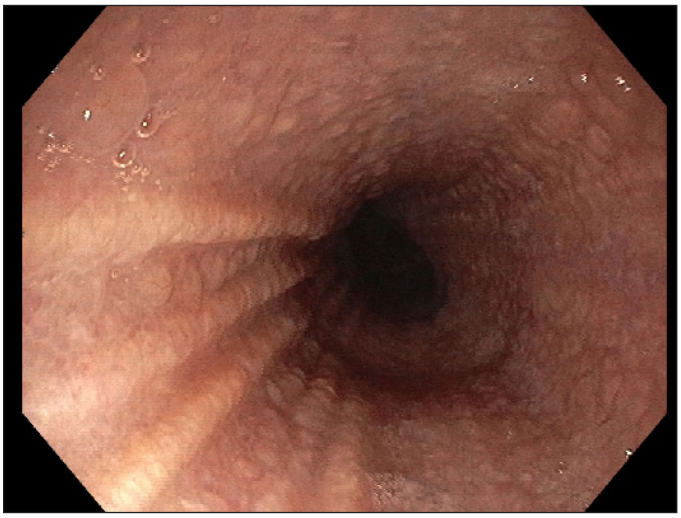

Figure 1. ***Endoscopic view of the proximal oesophagus, showing granular esophageal mucosa.***

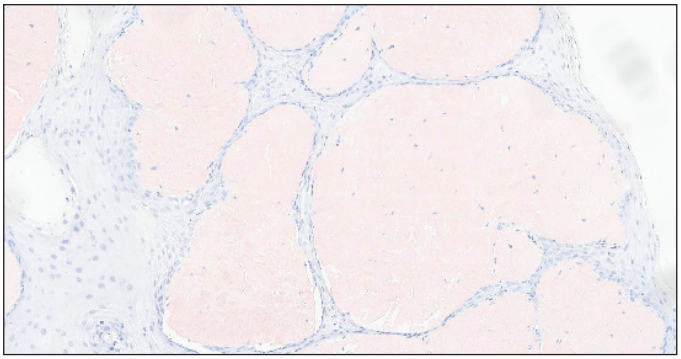

Figure 2. ***Visualisation of amyloid deposits by Congo Red staining.***

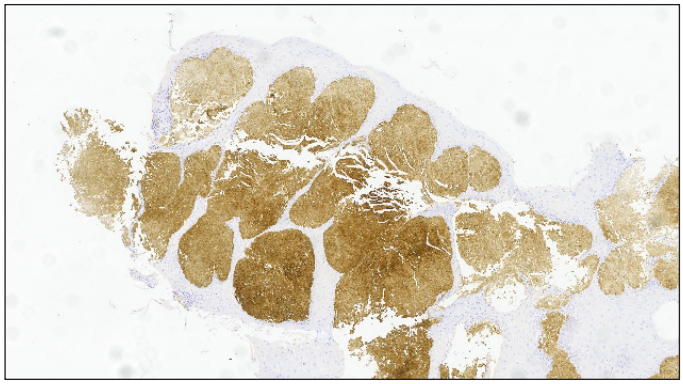

Figure 3. ***Applying polarised light to the tissue section shows apple-green birefringence.***

Discussion

Esophageal amyloidosis is usually silent and, if symptomatic, gastro-esophageal reflux is the most common clinical manifestation. Dysphagia, on the other hand, is a rare entity in this context. Endoscopically, mucosal friability, erosions, ulcers and submucosal haematomas are usually observed. Given the variability of endoscopic findings, as well as the presentation of non-specific symptoms, confirmatory diagnosis requires histopathological studies such as Congo red staining or birefringence under polarised light.

The mechanism of dysphagia secondary to esophageal amyloidosis is unknown, although part of it is attributed to a certain component of dysmotility secondary to atrophy due to nerve damage and pressure generated by the amyloid deposit as it settles between the muscle fibres. For this reason, the use of functional tests such as barium transit or esophageal manometry are also of interest, as they can offer characteristic radiological patterns in some cases (esophageal dilatation with distal narrowing)[1,2].

To date, there are hardly any described cases of dysphagia as the main manifestation of this disease, so examples such as this one should help us not to forget this diagnostic possibility, integrating esophageal amyloidosis into the differential diagnoses of dysphagia.

Bibliography

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