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European Journal of Radiology 63 (2007) 105–109

EJR
EUROPEAN JOURNAL OF RADIOLOGY

www.elsevier.com/locate/ejrad

Amyloidosis of the small intestine

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Received 13 January 2007; accepted 19 January 2007

Abstract

Amyloidosis is a rare disease characterized by forming pathological protein deposits – amyloid – in many organs and tissues. This decreases their functionality. The aim of this small study was to determine, whether the radiological picture of the small intestine involvement in amyloidosis is in some sense specific as sometimes described in literature giving rise to high suspicion for the disease in symptomatic patients.

Material and methods: The prospective study comprising seven patients hospitalized in surgical department is presented together with a survey on the disease, its appearance in radiological imaging. All patients underwent abdominal ultrasound (ATL 5000 HDI, 7–12 MHz linear probe, no contrast enhancement, supine position), abdominal CT (Somatom Plus, Siemens, single detector, conventional abdominal CT protocol) and enteroclysis (Micropaque suspension 300 ml, application rate of 75 ml/min, dilution with HP-7000 being 1:1 and HP-7000 solution 2000 ml, application rate of 120 ml/min.).

Results: The amyloid deposits in the small intestine could be visualized in five of seven patients with the disease. Enteroclysis revealed a diffuse slowed down intestinal motility with an obstruction-like picture in all of our seven patients. The intestinal secretion was normal, plicae were getting polyp-like shape in five of them forming so called "thumb printing" picture. CT showed thickening of the intestinal wall due to deposits with poor blood supply and contrast retention in five of seven patients. Ultrasound visualized thickened, hypoechoic nodular plicae and slowed down motility in these five patients. The most striking finding was the pathological deposits in the intestinal wall were highly hypo-vascular. However, this picture is very similar to that of ischemic enteritis. All seven patients had proven amyloid deposits from bioptic specimens.

Conclusion: The diagnosis of amyloidosis must be supported by bioptic examination as it has no pathognomic radiological picture. Nevertheless, it allows usually to rise a high suspicion for this and sometimes even guess the type of the pathological protein involved. This can start a search for the primary reason of possible amyloidosis and thus perhaps spear the patients quality of life.

Keywords: Small intestine; Tumor; Amyloidosis; Enteroclysis; Ultrasound; Ischemic enteritis

1. Introduction

Amyloidosis is a heterogeneous group of relatively rare impairments that all lead to a formation of hyaline deposits, i.e. extracellular foci of insoluble protein-mucopolysaccharide complex. The ground of the deposit formation is not clear yet. Nevertheless, it is presumed that prolonged antigenic stimulation shifts immunoregulation somehow. The disease does not usually spear an organ. Only 20% of the patients are suffering from a localised disease. Up to 15% of the patients with myeloma are forming the deposits of amyloid. Three clinical types according to presumed origin of amyloidosis could be distinguished—primary, secondary (in response to another disease) and senile [1].

The primary amyloidosis affects both parenchymatous organs and the digestive tube. Its origin stays hidden and men are usually getting ill more frequently. Mean age of the patients varies around 55–60 years. Renal insufficiency is a common sign. Heart, tongue, skin, kidney, nerves and the digestive tract are usually being affected. Patients are dying of renal failure or heart congestion [1]. The primary amyloidosis has a familiar predisposition very often accompanied by the monoclonal gammapathy.

The secondary amyloid is produced during several long-lasting diseases or when the organism is somehow irritated for a long time (chronic inflammation, etc.). The pathological sub-

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doi:10.1016/j.ejrad.2007.01.024

stance tends to form deposits in parenchymatous organs of the abdominal cavity and in lymph nodes.

The senile amyloidosis is usually manifesting in patients of higher age. This amyloid invades the brain, cardiovascular system, seminal vesicles and Langerhans' islands.

Clinical symptoms depend on the actual location of the deposits. Diarrhoea as well as intestinal obstruction, haemorrhage, malabsorption, infarction and perforation of the digestive tube may happen.

A biopsy specimen taken from the wall of the small intestine proves the diagnosis only in 70%. Besides that, rectal or gingival biopsy could be chosen as well too. The amyloid deposits can be found along small vessels in the submucosa and within their wall (beneath their intima as a cell-free deposit). But amyloid forms deposits within the smooth muscle fibres of the wall and in neural plexus too (it is typical for hereditary Portuguese or Iowa form and typically beta-2-microglobulines). The primary amyloidosis is often referred to as the light amyloid amyloidosis. It accompanies monoclonal proliferation of B cells producing immunoglobulins (mainly plasmocytes – multiple myeloma, Waldenstrőm's macroglobulinaemia and others belong here, for example) where the amyloid is made up of the light chains of immunoglobulins or their fragments. The secondary – AA – amyloidosis, the source of which lies in an increased release of one of the acute phase proteins – so-called SAA (serum amyloid-associated protein) from the liver and its following inappropriate proteolysis. This type of amyloidosis occurs even in tumorous decaying processes. Amyloid must be distinguished from hyaline – a product of regressive processes. Because it is impossible to be done by ordinary dyeing techniques, some special ones or electron microscopy must be engaged.

2. Materials and methods

Imaging findings and clinical charts of seven patients admitted to our surgical department during the period 2000–2005 suffering from a severe dyspepsia arising from intestinal amyloidosis have been considered in our prospective study. We have tried to find, whether there is a difference in sensitivity of three typical methods used for the small intestine imaging – CT, US and enteroclysis, and whether all these patients would present typical amyloidosis picture. All three methods were applied to all these seven patients within the small time range of 1 month from their admission.

The abdominal ultrasound targeted to the small intestine was performed my means of ATL 5000 HDI machine with the 7–12 MHz linear probe. No contrast enhancement was used. The examinations were performed without any preparation of the patients. The patients were examined in the supine position. First, the terminal ileum (the wall thickness, vascularization, echogenity of the surrounding fat, presence of nodes and course of peristalsis), caecum or the areas of preceding anastomoses were examined. The colon was assessed only superficially to get an overview (haustration, thickening of the wall). The intestinal folds morphology, thickness of its wall, vascularization of the thickened wall and the course of peristalsis were observed in the jejunum and ileum. Assessing the mesentery (presence and size of nodes, their shape, echogeneity and vascularization) and measuring the flows in AMS completed the examination. Our conventional abdominal CT protocol was used to visualize the small intestine pathology. Somatom Plus by Siemens, single detector, was used for this purpose. Native and i.v. contrast scans were performed after oral fractionated contrast enema administration, the last 500 ml right before the very examination. The area evaluated ranged from symphysis to the diaphragm, slice thickness 8 mm, table shift 12 mm. All patients underwent enteroclysis with Micropaque suspension 300 ml with the application rate of 75 ml/min, diluted with HP-7000 being 1:1 followed by HP-7000 solution 2000 ml with the application rate of 120 ml/min. Contrast media were applied by an enteroclysis catheter inserted under the topic local anesthesia through the nose. The application rate of the contrast medium was adjusted by means of an automatic pump. Less or more typical findings were searched for. Amongst them the most typical were obstruction-like picture with no real tumor, thickening of the wall with a poor blood supply, smooth mucosal surface, widespread and homogeneous intestinal affections, thickening of the plicae and the wall itself, intestinal dilation.

3. Results

All seven patients were admitted in our department because of fear of an acute surgery due to their dyspepsia decompensation. As the amyloid deposits in the small intestine were revealed and their state got improved on a wedged parenteral nutrition with very soon return back to the normal diet, no surgery was needed in all of them. There was no blood nor other pathological findings in their stool. None of them required surgery. The diagnosis was proven in all the cases by histological specimen taken by the upper or lower gastrointestinal endoscopy.

Radiological findings were very similar in all these patients. Enteroclysis showed slowed down motility in all seven patients with nearly obstruction-like picture in patients with high degree dyspepsia. Normal intestinal secretion. Thickened intestinal folds so that they resembled polyps and thumb-prints were found in five of them (Figs. 1 and 2). Villi were atrophic. Smooth mucosa. No ulcerations. No defects of the contrast filling. Obstruction-like picture was present. There was no real obstruction, it was rather the obstruction-like picture caused by the amyloid deposits. The wall was thickened, mainly the submucosa. Surroundings lacked any pathology. Other changes visible were that the process was typically widespread – diffuse.

CT revealed hepatomegaly in two patients showing their liver be involved too. Both native and post-contrast pictures showed focal areas of lower density. The liver was in six of seven patients inhomogeneous, of a marble-like structure. Lower density areas representing the deposits of amyloid retained more or less contrast in the post-contrast phase of the examination. Amyloid was seen mainly in the parenchyma, along sinusoids in two patients what is common in primary amyloid. Four patients had different amyloid distribution within the wall of vessels what may be typical for the secondary amyloid. A rare local accumulation of amyloid happened in one of these patients so that it formed tumour-like lesions that were hypodense with poor blood supply