Amyloid tumour of the colon

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Accepted for publication
13 October 1994

Abstract

A case of amyloid tumour of the colon and the first in association with a carcinoma is reported. A previously healthy 65 year old man presented with non-specific symptoms of lower abdominal pain and flatulence without rectal bleeding. A clinical diagnosis of diverticular disease was made and colonoscopy performed. Two lesions (one at 15 cm and the other at 30 cm from the anal margin) were found on endoscopy and removed. On histology, the lesion at 15 cm was a moderately differentiated adenocarcinoma and that at 30 cm contained amyloid. Further tests (standard tinctorial methods and immunohistochemistry) revealed the 30 cm lesion to be an amyloid tumour of the colon of AL (λ) type. When biopsy of an atypical,

large, solitary colorectal lesion reveals amyloid deposition, the possibility of an amyloid tumour should be considered and the lesion resected.

(*J Clin Pathol* 1995;48:592-593)

Keywords: Amyloid tumour, AL-amyloid, colon, colorectal carcinoma.

Diffuse deposition of amyloid in the rectum is a well recognised occurrence in many chronic systemic diseases. Rarely, large localised collections of amyloid ("amyloid tumours") may develop in the absence of underlying chronic disease. Although these may arise at several sites,¹⁻⁵ only one previous case has been reported in the rectum.⁶ We report a further case of amyloid tumour of the colon and the first in association with a carcinoma.

Case report

A previously healthy 65 year old man was referred with non-specific symptoms of lower abdominal pain and flatulence without rectal bleeding. A clinical diagnosis of diverticular disease was made and colonoscopy performed. Endoscopy revealed two polypoid lesions: one a villous lesion at 15 cm and the other a large, dark brown, haemorrhagic, sessile lesion 30 cm from the anal margin. Biopsy suggested the lesion at 15 cm to be a tubulo-villous adenoma with moderate dysplasia and the lesion at 30 cm to contain amyloid. At laparotomy, no other pathology was apparent and both lesions were removed by an uneventful anterior resection of the rectum and sigmoid colectomy (fig 1).

Histopathological examination revealed the lesion at 15 cm to be a moderately differentiated adenocarcinoma arising in a tubulo-villous adenoma with one of six recovered lymph nodes involved by metastatic carcinoma. The lesion at 30 cm measured 7 × 6 cm in surface area and was well circumscribed with a blue-purple external appearance. This lesion was composed of a submucosal deposit of amyloid, both lying free in the submucosa and involving submucosal vessels, with attenuation and regeneration of the overlying mucosa. There was extensive haemorrhage with the submucosa. In places, the amyloid deposit elicited a foreign body giant cell response (fig 2). Congo Red staining was positive, both before and after pretreatment with potassium permanganate and revealed apple-green birefringence under cross-polarised light. Immunohistochemical staining was negative for serum amyloid A protein, prealbumin (transthyretin) and κ chains, but was positive for λ chains. These

Figure 1 Gross photograph showing adenocarcinoma arising in a tubulovillous adenoma (below) and solitary amyloid tumour (above).

Figure 2 Higher power view stained with Congo Red showing vessel wall and free submucosal amyloid involvement. Note foreign body giant cell response (Congo Red, $\times 400$)

were the appearances of an amyloid tumour of the rectum of AL (λ) type. Subsequent clinical investigations, including serum and urine electrophoresis and bone marrow studies, were negative. The patient is currently asymptomatic 12 months after surgery.

Discussion

In all forms of amyloidosis the gastrointestinal tract is involved in between 70 and 100% of cases.⁷ Rectal biopsy is the recognised technique for confirming a diagnosis of systemic amyloidosis. Isolated masses of amyloid deposition ("amyloid tumours") without systemic involvement are rare but well documented and have been found in a number of sites such as the oesophagus,¹ lung² and breast.³ To the best of our knowledge, there has been one previous case of "amyloid tumour" of the rectum⁶ but this was not associated with a synchronous carcinoma. However, colonic carcinoma has been reported with colonic amyloidosis⁸ but not an amyloid tumour. This may be the first case of a large colonic "amyloid tumour" in association with a rectal carcinoma.

The aetiology of amyloid tumours is uncertain. Some cases have been attributed to

chronic inflammation such as gonococcal urethritis, osteoarthritic joints and damaged heart valves,^{6,7} presumably related to AA-amyloid. A form of dystrophic amyloid deposition has been described in areas of chronic tissue damage.⁵ In our case standard tinctorial methods and immunohistochemistry suggested that the amyloid deposited was of AL-type, probably of λ light chain origin. Investigations did not reveal the source of this protein and no other localised or generalised colorectal condition—for example, inflammatory bowel disease, was apparent. The synchronous adenocarcinoma was unlikely to have been contributory, being sited a considerable distance from the amyloid tumour. Amyloid usually associated with neoplasia, such as renal cell carcinoma and Hodgkin's disease, is of AA-type and neither tumour type has been reported in association with solitary amyloid tumours.

This patient presented with lower abdominal pain and flatulence, and such symptoms may be attributable to "amyloid tumour" or adenocarcinoma. It is interesting to note that involvement of intestinal vessels by β_2 -microglobulin associated amyloidosis in patients undergoing long term haemodialysis may lead to ischaemic ulceration⁹ and infarction,¹⁰ and the haemorrhagic nature of the submucosa and regenerative features noted in the attenuated overlying mucosa in this case may represent a response to low grade ischaemia. Amyloid tumours at other sites tend to run a benign course with resection being curative.¹² This is likely to be the case in colonic amyloid tumours and the prognosis in the present case will probably be determined by the behaviour of the synchronous rectal adenocarcinoma.

In conclusion, when biopsy of an atypical, large, solitary lesion in the colorectum reveals amyloid deposition, the possibility of an amyloid tumour should be considered and the lesion resected.

We wish to thank Mrs C Bartley for performing the tinctorial and immunohistochemical stains and Mrs J Crosby for photographic assistance.

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