Elastofibromatous polyp of the sigmoid colon – A case report and review of gastrointestinal elastofibromas

Z Vesoulis MD¹, Pars Ravichandran MD¹, Dimitri Agamanolis MD², Duane Roe MD³

Z Vesoulis, P Ravichandran, D Agamanolis, D Roe. Elastofibromatous polyp of the sigmoid colon – A case report and review of gastrointestinal elastofibromas. *Can J Gastroenterol* 2003;17(4):275-277.

Elastofibromatous change in the gastrointestinal tract is a rarely reported, usually polypoid lesion of unknown etiology with submucosal stromal change that may mimic amyloid deposition. The constituent amorphous material of the polyp stroma has distinctive features that permit an accurate assessment and diagnosis including: distribution of the material predominantly in the submucosa; distinctive fibrillar and granular appearance of the deposits; thick, irregular, haphazardly arranged bundles of elastic fibres positive for Verhoeff's elastic stain; ultrastructural fibres with an electron dense curvilinear or beaded appearance; lack of amyloid type vascular wall deposits; and lack of amyloid congophilia or crystal violet metachromasia. The clinical, light microscopic, histochemical and ultrastructural characteristics of this deposited material are reviewed in detail in the present report of a patient who presented with an asymptomatic polypoid lesion of the sigmoid colon. Other reported cases are summarized, and their clinical and pathological features are compared with the current case.

Key Words: *Gastrointestinal amyloid; Granulofibrillar deposits; Polypoid elastofibroma*

Polype élastofibromateux du sigmoïde – Rapport de cas et revue des élastofibromes gastro-intestinaux

Un changement élastofibromateux dans l'appareil gastro-intestinal est une lésion rarement rapportée, habituellement de type polypoidé d'étiologie inconnue présentant une modification du stroma sous-muqueux et s'apparentant à une accumulation de substance amyloïde. La matière amorphe qui constitue le stroma du polype possède des caractéristiques distinctives qui permettent une évaluation et un diagnostic précis incluant : la distribution de la matière surtout dans la sous-muqueuse; l'apparence granuleuse et fibreuse distinctive des dépôts; les amas épais, irréguliers et désordonnés de fibres élastiques positives à la coloration de Verhoeff; les fibres ultrastructurelles d'apparence curviligne ou perlée dense aux électrons; l'absence de dépôts de type amyloïde dans la paroi vasculaire; et l'absence de congophilie amyloïde ou de métachromasie au cristal violet. Les caractéristiques ultrastructurelles, histochimiques, de microscopie optique et cliniques de ces dépôts sont étudiées en détail dans la présente observation d'un patient présentant une lésion polypoidé asymptomatique du sigmoïde. D'autres cas sont également résumés et leurs caractéristiques pathologiques et cliniques sont comparées à celle du cas à l'étude.

Elastofibromatous lesions are most commonly found as soft tissue masses. The prototype elastofibroma dorsi, a relatively uncommon neoplasm-like lesion, usually occurs in the subscapular region of older adults and is thought to be a degenerative phenomenon secondary to repetitive use trauma. Elastofibromas have uncommonly been described in the hand, foot, eye, mediastinum and omentum (1-6). Accounts of gastrointestinal tract elastofibromatous lesions have been both rare and pathologically variable, reported most often in Asian patients, with lesions of the stomach, sigmoid colon and rectum (7-10). Elastofibromatous lesions have a distinctive histology that includes the presence of broad, fragmented elastic fibres that may be readily identified on hematoxylin and eosin stains (H&E) but are highlighted with the Verhoeff van Gieson (EVG) and Verhoeff iron hematoxylin staining. Some reports have described a close H&E morphological similarity of this elastic-rich material to amyloid an important histological distinction with significant clinical implications.

CASE REPORT

A 72-year-old white male patient was referred to the gastroenterology service at Summa Health Systems (Akron, Ohio) subsequent to the discovery of a rectosigmoid polyp on a routine screening sigmoidoscopy at another institution. His past medical history included one episode of transient ischemic attack in the previous year and a cholecystectomy several years before. There was no history of upper or lower gastrointestinal tract bleeding. His current medications included Zocor (Merck Frosst, Kirkland, Quebec), Plavix (Bristol-Myers Squibb/Sanofi Canada, Markham, Ontario), and over the counter vitamin supplements.

Using standard protocol, a full colonoscopy was performed. The colonoscope was advanced without difficulty to the cecum, and the ileocecal valve was clearly visualized. On withdrawing the instrument, an 8 mm umbilicated, slightly elongated, smoothly outlined sessile polyp that had a cream to yellow colouration was seen in the sigmoid colon (Figure 1). The gastroenterologist thought this lesion closely resembled a submucosal lipoma. Using

¹Department of Pathology, Summa Health System, Akron City Hospital; ²Department of Pathology, Akron Children's Hospital; ³Department of Gastroenterology, Summa Health System, Akron City Hospital, Akron, Ohio, USA

Correspondence and reprints: Dr Z Vesoulis, Department of Pathology, Akron City Hospital, 525 East Market Street, Akron, Ohio 44304, USA
Telephone 330-375-3902, fax 330-375-4874, e-mail vesouliz@summa-health.org

Received for publication October 31, 2002. Accepted February 12, 2003

Figure 1) Endoscopic appearance of a sigmoid polypoid elastofibroma showing a 0.8 cm smooth, slightly elongated sessile polyp, somewhat resembling a gastrointestinal lipoma (arrow)

Figure 2) The polypoid sigmoid lesion shows a core of eosinophilic granulofibrillar amorphous material mostly confined to the submucosa and distinct separation from the lamina propria (Hematoxylin and eosin stain, original magnification $\times 100$)

an electrocautery snare, the polyp was completely removed without significant bleeding or other complications. No additional lesions were discovered in the remainder of the colon.

METHODS

Sigmoid colon biopsy tissue obtained during the colonoscopy was received in 10% formalin and processed for histology in the usual manner. Sections were stained with H&E, EVG, Congo red and crystal violet. For ultrastructural analysis, formalin-fixed tumour tissue was removed from a paraffin block, deparaffinized and transferred to 3% glutaraldehyde. This material was subsequently fixed in an epoxy resin. Thin plastic sections were stained with uranyl acetate and lead citrate. These sections were evaluated using electron microscopy (Zeiss model 109, Oberkochen, Germany).

PATHOLOGY

A 0.8 cm polypoid lesion of the sigmoid colon demonstrated a cut surface with a homogeneous gray-yellow colouration and a soft rubber-like consistency.

Figure 3) Amorphous submucosal material nearly obliterates the muscularis mucosa with splaying of the residual fibres of the muscularis (arrow). (Hematoxylin and eosin stain, original magnification $\times 200$)

Figure 4) Elastic stain of the submucosal deposits demonstrating numerous randomly distributed, broad and fragmented elastic fibres embedded in a fibrillar and granular matrix. (Verhoeff van Gieson stain, original magnification $\times 400$)

Histologically, the mucosa was unremarkable, but the submucosa was diffusely replaced by a rounded mass of homogeneous granular to fibrillar-appearing eosinophilic material (Figure 2) with splaying of the fibres of the muscularis mucosia (Figure 3). This material did not have the opaque 'fluffy' or 'cracked' appearance of typical amyloid and the distribution of the amorphous material seemed inconsistent with amyloid. There were no significant lamina propria infiltrates or restriction of deposits to submucosal vessels, more typical of gastrointestinal amyloid deposition. Submucosal vessels appeared histologically unremarkable. Special stains for amyloid including Congo red and crystal violet lacked congophilia, apple-green birefringence or metachromasia. Elastic staining (EVG) was positive for thick, truncated and randomly distributed elastic fibres embedded in the granular matrix (Figure 4). Paraffin-embedded tissue from the polyp was also examined by

TABLE 1
Reported gastrointestinal elastofibromas

Site (reference)	Polypoid	Distribution of elastic fibres	Elastic stain	Amyloid stains	Age/Sex	History	Elastofibroma dorsi	Lesion size
Sigmoid (present case)	Yes	Submucosa	EVG +	CR, CV-	72/M	Asymptomatic	No	0.8 cm
Rectum (8)	No	Submucosa	EVG +	CR-	58/F	Myeloma	No	NR
Stomach (7)	No	Submucosa	Elastic +	NR	69/F	Peptic ulcer	Yes	6 × 8 cm
Colon (transverse) (9)	Yes	Submucosa	Elastic +	NR	49/M	Colon adenoma	No	0.5 cm
Sigmoid (10)	Yes	Submucosa	Elastic +	NR	69/F	Colon adenoma	No	0.7 cm

CR Congo red; CV Crystal violet; EVG Verhoeff van Gieson; F Female; M Male NR Not reported

electron microscopy. Ultrastructurally, the granulofibrillar material was predominantly composed of collagen fibres, with scattered electron dense, coarse curvilinear and beaded fibres consistent with elastin. Rare oval and spindle shaped nuclei of stromal cells were also seen. There was no ultrastructural evidence of non-branching fibres typical of amyloid.

DISCUSSION

When arising in soft tissue, elastofibromas are probably not true neoplasms. These tumour-like growths are slow growing, ill defined masses in older individuals, often with an occupational association involving strenuous manual labour. Typically, these masses are large, reaching 5 cm to 10 cm in size and are found in a consistent location between the lower scapula and chest wall. Gastrointestinal elastofibromatous change is a lesion of unknown etiology, sometimes attributed to a postinflammatory or post-traumatic condition (6,7,10), somewhat similar to its soft tissue histogenesis. Most have been reported as small polypoid lesions of the colon. The inciting 'traumatic' incident of their genesis is undefined. A postinflammatory origin seems unlikely, because none of these lesions has ever been associated with any form of specific or idiopathic inflammatory bowel disease. Gastrointestinal elastofibromatous change may be a relatively uncommon and atypical reparative process, because some ultrastructural studies have suggested an overproduction of elastic fibres by fibroblasts and reactive elastin synthesis (11,12).

The current report and at least one previous report indicate the potential differential diagnostic confusion with gastrointestinal amyloid (8). Clinically, the patient with amyloid may have chronic inflammatory or degenerative diseases, a plasma cell dyscrasia, monoclonal gammopathy, or evidence of systemic or multiorgan involvement. Endoscopically, the typical appearance of gastrointestinal amyloid includes granular mucosa, friable hemorrhagic

mucosal plaques or thickened folds (13). The typical elastofibroma appears usually as an incidental, innocuous appearing, solitary polypoid lesion in the asymptomatic patient. Histologically, accumulations of amyloid occur most commonly as thick, heterogeneous, opaque, fluffy deposits in the walls of small submucosal vessels. Uncommonly, the amyloid deposits may appear as amorphous, eosinophilic coalescent globules in the lamina propria. In familial Mediterranean fever, amyloid deposits may diffusely infiltrate the muscularis propria and the myenteric plexus of the small bowel with consequent hypomotility, bacterial overgrowth and malabsorption (14). Elastofibromatous lesions most commonly occur as polyps of the rectosigmoid region with a distinctive deposition of granulofibrillar material reported exclusively in the submucosa (Table 1). A distinction between elastofibromatous mucosal change and amyloid deposition also can be readily made after examination of slides stained with elastic and amyloid histochemical stains. Elastic stains are uniformly positive for thick dense fibres and the positive staining disappears with elastase digestion in some reported cases in which digestion techniques were used (9,10). Rarely is ultrastructural confirmation or immunohistochemical positive staining with CD34 necessary (15).

Gastrointestinal elastofibromas appear to occur on a sporadic basis almost exclusively in the colon with a rare gastric occurrence (7). Patients with these lesions have clinically presented with varied symptoms and findings that appear to be unrelated to their gastrointestinal elastofibroma. Only few reports have attributed any symptomatology directly to the elastofibroma, with the majority presenting incidentally at endoscopy (Table 1). The primary significance of the gastrointestinal elastofibroma lies in its appropriate recognition as a rare benign polypoid lesion of the colonic mucosa, its distinction from other benign stromal polyps, particularly lipomas or hyperplastic polyps, and its differentiation from gastrointestinal amyloid deposition.

REFERENCES

1. Austin P, Jakobiec FA, Iwamoto T, Hornblase A. Elastofibroma oculi. Arch Ophthalmol 1983;101:1575-9.
2. Cross DL, Mills SE, Kulund DN. Elastofibroma arising in the foot. South Med J 1984;77:1194-6.
3. De Nicolis M, Goteri G, Campanati G, Prat J. Elastofibrolipoma of the mediastinum. A previously undescribed benign tumor containing abnormal elastic fibers. Am J Surg Pathol 1995;19:364-7.
4. Hsu JK, Cavanagh HD, Green WR. An unusual case of elastofibroma oculi. Cornea 1997;16:112-9.
5. Kapff PD, Hocken DB, Simpson RH. Elastofibroma of the hand. J Bone Joint Surg Br 1987;69:468-9.
6. Tsutsumi A, Kawabata K, Taguchi K, Doi K. Elastofibroma of the greater omentum. Acta Pathol Jpn 1985;35:233-41.
7. Enjoji M, Sumiyoshi K, Sueyoshi K. Elastofibromatous lesion of the stomach in a patient with elastofibroma dorsi. Am J Surg Pathol 1985;9:233-7.
8. Goldblum JR, Beals T, Weiss SW. Elastofibromatous change of the rectum. A lesion mimicking amyloidosis. Am J Surg Pathol 1992;16:793-5.
9. Hayashi K, Ohtsuki Y, Sonobe H, et al. Pre-elastofibroma-like colonic polyp: Another cause of colonic polyp. Acta Med Okayama 1991;45:49-53.
10. Sakatani T, Shomori K, Adachi H, Hosoda A, Ito H. Elastofibroma of the sigmoid colon. Pathol Res Pract 2000;196:205-7.
11. Dixon AY, Lee SH. An ultrastructural study of elastofibromas. Hum Pathol 1980;11:257-62.
12. Kindblom LG, Spicer SS. Elastofibroma. A correlated light and electron microscopic study. Virchows Arch A Pathol Anat Histol 1982;396:127-40.
13. Tada S, Iida M, Yao T, et al. Endoscopic features in amyloidosis of the small intestine: Clinical and morphologic differences between chemical types of amyloid protein. Gastrointest Endosc 1994;40:45-50.
14. Ravid M, Sohar E. Intestinal malabsorption: First manifestation of amyloidosis in familial Mediterranean fever. Gastroenterol 1974;66:446-9.
15. Ruelas Villavicencio AL, Aponte Paredes PS, Ortiz Hidalgo C. CD-34 expression in elastofibroma. Clinicopathological, histochemical, and immunohistochemical study of four cases. Rev Invest Clin 1999;51:11-6.