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

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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

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-Meyers 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 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.
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.

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 mucosa (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

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

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

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 post-inflammatory or post-traumatic condition (6,7,10), somewhat similar to its soft tissue histogenensis. Most have been reported as small polypoid lesions of the colon. The inciting 'traumatic' incident of their genesis is undefined. A post-inflammation 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

