Review

Elastotic Changes in the Gastrointestinal Tract: A Review of Literature

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Elastotic changes are benign lesions in the gastrointestinal tract that often appear as polyps and show histologically a remarkable increase in elastic fibers. Because of their hyaline and amorphous appearance in hematoxylin and eosin (H&E) stained slides, elastotic changes often resemble amyloidosis. However, they are negative in Congo red staining. Reviewing the literature we found 35 cases in 12 publications in the English and French literature since 1985. The results indicate that the patients' age ranged from 24 to 88 years (mean 58.2, median 58) and presented a balanced gender ratio (M/F = 17/18). Usually the lesions presented as polyps or irregular mucosal areas. Mostly, they were found during endoscopic examination in the colon or rectum (16 cases), while six cases were located in the stomach and only two in the small bowel. Some authors consider the alterations to be a reactive process, e.g. within (gastric) ulcers, whereas others speculate about a connection with systemic diseases of the connective tissue (*Ehlers-Danlos Syndrome, Pseudoxanthoma elasticum*). Based on our observations we distinguished *angioelastosis,* showing a relation to submucosal vessels, as a separate entity from *gastrointestinal elastofibroma,* that presents the same histologic morphology as *Elastofibroma dorsi.* Conclusion: Elastofibromatous changes in the gastrointestinal tract are quite common benign findings and should be considered as a possible differential diagnosis in examining gastrointestinal specimens.
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Key Words: *gastrointestinal tract, polyps, elastosis, angioelastosis, elastofibroma*

INTRODUCTION

Elastofibroma dorsi was first described in 1961 by Järvi and Saxén[1] as a fibrous lesion with abnormal elastic fibers located mostly in subscapular soft tissue. In other anatomic regions -especially in the gastrointestinal tract- elastotic changes were found in different locations, including the stomach, small bowel, colon and rectum (see **Table 1**). In general however, gastrointestinal lesions found only minor reflection in the literature with single cases reports and small series. Depending on the authors, these changes were termed *elastofibroma*, *pre-elastofibroma-like lesion*, *colonic elastoma*, *elastosis* or *gastrointestinal angioelastosis.*

The cases mostly appeared as polyps or polypoid changes in endoscopic or macroscopic examination, sometimes with surface ulceration. One case is described with no gross alterations, but was just a coincident finding after rectal biopsy.[2] Because elastotic changes in the gastrointestinal tract (GIT) are not described in common textbooks they seem to be widely unknown and are not generally accepted as own entity. This, however, may cause problems in evaluating such specimens which are identifyed by the clinicians and do not fit into conventional categories of polyp causing lesions.

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Histologically, all of these changes had submucosal and or mucosal deposits of increased elastic fibers that appeared amorphous and eosinophilic in H&E stained slides and, thus, amyloid-like. However, when Congo red staining was performed, the results were always negative, in contrast to elastic staining, which highlighted the elastic fibers. In some of the cases, relations to submucosal blood vessels are described.

METHODS

We performed a literature search within the Medline and Google Scholar data bases using combinations of the terms elastosis, elastotic, gastrointestinal, colon, rectum, colorectal and stomach, esophagus. The identified publications were screened for relevance and the reference lists were checked concerning publications that were not included yet. The search period was not restricted.

It is the goal of this review to describe the spectrum of elastotic changes in the GIT which is an alteration that did not find recognition in textbooks and can therefore cause diagnostic problems.

MORPHOLOGY

In 1985, Enjoji et al. were the first to publish a case report about an elastofibromatous lesion in the stomach (see **Table 1**). The female patient underwent partial gastrectomy because of a gastric ulcer and grossly showed a wide thickening of the

antral wall with rubbery elastic consistency.[3] There are 5 further cases that report similar changes in the stomach, mostly in the antrum.[4-7] One of these cases also had a gastric ulcer,[6] others showed erosion or inflammatory changes,[4,7] two patients were suspected to have gastric cancer[5,7] and another two had undergone a Billroth-operation.[4] Only one of the gastric cases refers to evidence of bilateral subscapular *Elastofibroma dorsi*.[3]

Table 1. Data of 35 published cases of elastofibromatous changes in the gastrointestinal tract.

Authors	**Cases**	**Sex(F/M)/Age (years)**	**Location**
Enjoji et al. 1985	1	F/69	Stomach/Antrum
Hayashi et al. 1991	1	M/49	Colon/Transversum
Goldblum et al. 1992	1	F/58	Rectum
Kharsa et al. 1992	7	M/55	Colon
		M/34	Colon/Descendens
		F/30	Colon/Sigma
		M/32	Rectum
		F/58	Colon/Descendens
		F/35	Colon/Sigma
		M/46	Colon/Sigma
Schiffman 1993	1	F/88	Small Bowel
Sakatani et al. 2000	1	F/69	Colon/Sigma
Vesoulis et al. 2003	1	M/72	Colon/Sigma
Saint-Paul et al. 2003	1	F/76	Stomach/Antrum
Hobbs et al. 2004	13	F/63	Colon/70 cm
		M/53	Colon/25 cm
		F/61	Colon/Sigma
		M/77	Small Bowel/Ileum
		M/77	Stomach/Antrum
		F/51	Colon/Sigma
		F/71	Colon/Descendens
		M/83	Hepatic flexure
		M/55	Stomach
		F/61	Colon/Coecum
		M/47	Colon/50 cm
		F/50	Colon/Transversum
		M/47	Colon/Descendens
Märkl et al. 2008	6	M/45	Colon/Ascendens
		F/60	Colon/Sigma
		F/55	Rectum
		M/70	Colon/Left flexure
		M/24	Small Bowel/Ileum
		M/64	Colon/Descendens
Kai et al. 2009	1	F/77	Stomach/Antrum
Lau et al. 2010	1	F/75	Stomach/Antrum
	35	**Mean 58.2 y, Median 58 y**	

After the first report, others followed concerning elastofibromas or elastotoic changes not only in the stomach, but also in the colorectal region and in the small bowel. In the ileum, there are only three cases present, the first of which grossly appeared as a polyp.[8] The second patient suffered from small bowel obstruction with ulceration and hemorrhagic mucosa.[4] The third case was an incident finding in an autopsy case, which disclosed similar changes in the bronchial system.[9]

The remaining cases are colorectal biopsy or colectomy specimens with polypoid, sometimes cobblestone-like alterations.[2,4,8,10-13]

All 35 full published cases histologically had elastofibromatous characteristics in common: granular and/or fibrillar, eosinophilic, amorphous masses within connective submucosal tissue with scattered fibroblasts, small blood vessels and serrated outlines. Because these changes are highly suspicious for amyloidosis, Congo red staining was quite often performed, but always with negative results, as well as von Kossa staining. Elastic staining (Verhoeff's or EvG, respectively) showed consistent positivity of the amorphous material in all cases and identified it as elastic fibers.

Our group became the first to divide gastrointestinal elastosis into two main different groups or entities which are explained in the following.

Angioelastosis

We suggested the term angioelastosis for a lesions that shows an association between the densely accumulated elastic fibers and the wall of submucosal blood vessels.[8] Importantly, these elastic fibers have an untruncated structure and can reach the mucosal layer. Therefore, the vessel component can be missing in normal biopsies lacking submucosal structures (**Figure 1A**). In the literature we identified 12 cases according to the descriptions of the authors we probably would clasify as *angioelastosis*. We strongly believe that the vast majority of cases with elastotic changes belong to this group. All of them are located either in the colorectal region or rarely in the small bowel.

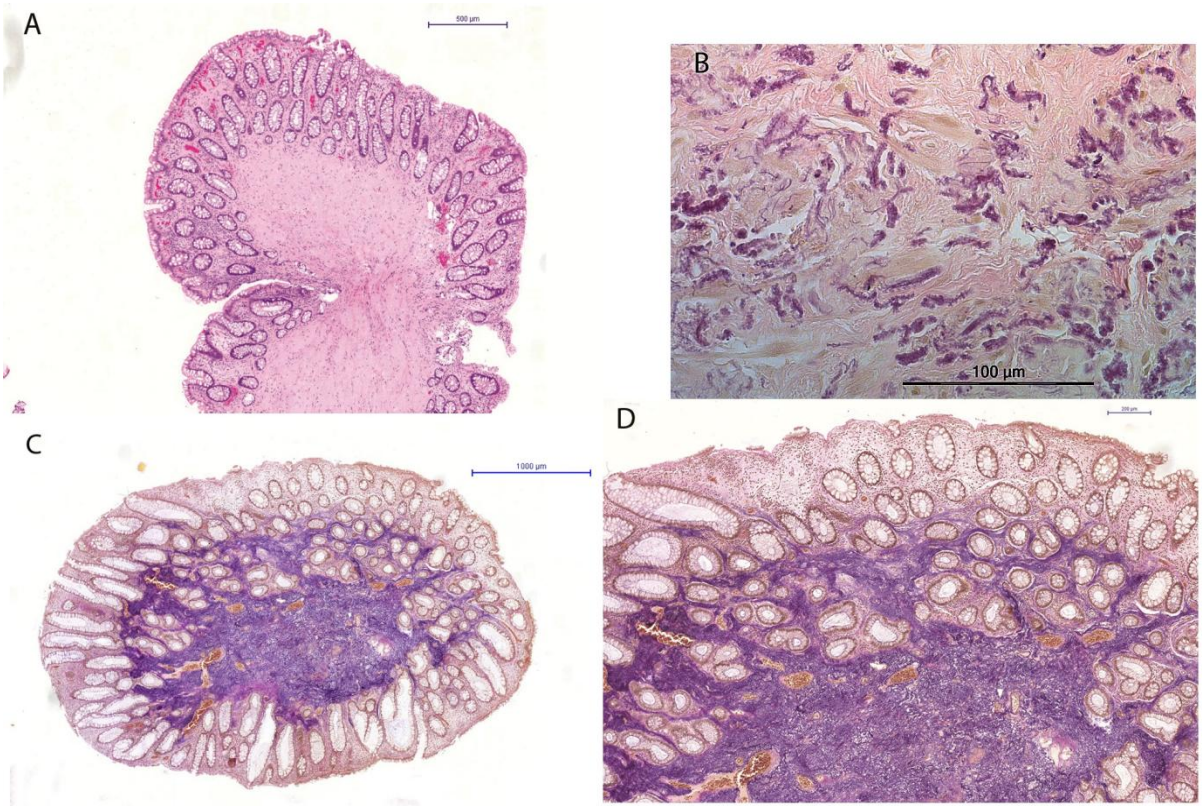

Figure 1. Specimens of colonic polyps.
A) H&E staining - Specimen of a colonic polyp with enlargement of the Lamina propria by an accumulation of fibroelastic fibers.
B) Elastica-van-Giesson-staining and higher magnification of A) - the elastic fibers show the typical truncated and globular morphology of an elastofibroma.
C) Elastica-van-Giesson-staining - Specimen of a rectum polyp with amorphous hyalinization of the Lamina propria.
D) Higher magnification of C) reveals a heavy accumulation of elastic fibers.

Elastofibroma

Lesions presenting with typical bended and truncated fibers with serrated outlines are probably true elastofibromas which are better known in the subscapular region. These alterations are typically located in the submucosal layer of the stomach causing a remarkable thickening (**Figure 1C**/**1D**). Only 3 cases (2 in the stomach, one in the sigma) have been published up to now.[3,7,12] We found 2 additional cases in our files (unpublished data). In 2009, Kai et al. published a case of an elastofibroma in the stomach with a dense accumulation of elastic fibres with a remarkable association to vessels at the border of the lesion. These latter lesion areas were termed *PVFLs (perivascular fibrotic lesions)* by the authors.[5]

It has to be stated that the well-known elastotic vessel alterations caused by neuroendocrine tunors (NET) do not fit into these categories and have to be separated from the lesions described in this review.

The Clinical Meaning of Elastotic Changes in the GIT

Based on unpublished data, we estimate the frequency of elastotic changes or *angioelastosis* to be about 1-2% of all incoming GIT biopsy specimens regardless of the indication for the endoscopy.Thus, it is a rare condition but not as exceedingly rare as originally thought. As mentioned before, these lesions often present as polyps or irregular mucosal areas. Neoplasia or even malignancy must be ruled out in such cases. Because the histological appearance is suggestive for amyloidosis Congo stain is performed often but turns out to be negative. As long as an elastic staining is not performed the cases remain unclear. On the other hand the endoscopic aspect can be very serious, prompting surveillance in short intervals because of unsatisfying histological findings. In contrast, elastotic changes are almost always associated with benign conditions. Previous endoscopic biopsies, previous radiation, atrophic gastritis and healed or healing ulcerations are the most frequent underlying findings. Therefore, angioelastosis seems to be a reactive process. In the opposite there is growing evidence that true elastofibromas are of neoplastic origin with benign behavior. Despite this usually benign nature, some elastotic lesions can cause severe stenosis, which lead to surgical resection to avoid complete stenosis and again to rule out a malignant process.

CONCLUSION

The number of published cases shows that elastofibromatous changes in the gastrointestinal tract are not as rare as originally thought. They appear in the stomach as wall thickening -e.g. related to ulceration or post-interventional status- as well as in small bowel, colon and rectum, often as polypoid lesions. As the elastic fibers build amorphous masses, the H&E staining gives the impression of amyloidosis. In order to diagnose correctly, a Congo staining should be performed to show a negative result, and an EvG staining should accentuate the increase of elastic fibers.

Even though the concrete cause of elastofibromatous changes remains unclear, authors agree that there is probably an association with alterations such as ulcers, inflammatory processes, radiotherapy or surgical treatment and, therefore, they can be seen as reactive lesions. Nevertheless, in some cases excessive elastotic changes are caused by a neoplastic process. It is well known that particular neuroendocrine tumors (NET) are often accompanied by remarkable elastotic changes. We found elastotic changes also in association with colonic adenoma (2 cases) and invasive colon cancer. The thesis by Kharsa et al., stating that systemic diseases of the connective tissue, such as *Ehlers-Danlos Syndrome* or *Pseudoxanthoma elasticum*, are the underlying cause could never (not even in their own study) be proved.[11] There might possibly be a relation to elastofibrosis in other locations in the same patient, but only two cases are known: one with changes in the small bowel and bronchus, and a single case report with changes in the stomach and 'classic' *Elastofibroma dorsi* located in the scapular region.[3,9]

We recently assigned two different categories of elastotic gastrointestinal changes, terming one of them *gastrointestinal angioelastosis* to confirm the involvement of submucosal blood vessels. The other one is thought to have exactly the same histological features as *Elastofibroma dorsi* and should consistently be termed *gastrointestinal elastofibroma*, analogous to the subscapular parent.

CONFLICT OF INTEREST

None.

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