Results of coagulation studies

	Pretreatment	Immediately after treatment	One month after treatment
Bleeding time (mins)	4.5		9
Prothrombin time (secs)	14	13	12
Control time (secs)	12	12	12
Kaolin-cephalin clotting time (KCCT) (secs)	72	40	42
Control time (secs)	40	41	47
KCCT corrections (secs)			
1 in 2 STAT	49		
30 mins	54		
Control STAT	40		
30 mins	44		
1 in 5 STAT	53		
30 mins	79		
Control STAT	41		
30 mins	44		
FVIII:c (%)	80		160
FIX:c (%)	90		110
FXI:c (%)	18	27	39
FXII:c (%)	65		44
FXI inhibitor (Bethesda Units)	0.8	0.4	<0.1

In patients with haemophilia who developed antibodies to factor VIII there is usually no detectable circulating factor VIII. In our patient there was a detectable concentration of factor XI in the plasma despite the presence of free antibody. It may be that the patient was producing sufficient factor XI to compensate for the destruction by the low inhibitor concentration, although the factor XI did not return to normal after the inhibitor became undetectable. Alternatively, it may be that the inhibitor required a cofactor which was depleted before the removal of all factor XI, although in this case an Exner test inhibitor pattern of type 3 would be expected.

In previous reports in which the acquired inhibitor has been further characterised it has been found to be of the IgG type.^{3,6} In our case incubation with anti-human IgG did not affect the factor XI inhibitor concentration, confirming that it was not an IgG type. Conversely, inhibitor activity was abolished by the addition

of 2-mercaptoethanol (2ME), which disrupts pentameric IgM, so the observed effect strongly suggests an IgM antibody. Unfortunately no further investigations such as incubation with anti-human IgM could be performed to confirm this.

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Solitary ganglioneuroma of the rectum: Report of two cases

T W Beer

Abstract

Two cases of solitary rectal ganglioneuromas are reported, one in a patient with several previously resected colorectal adenomas, the other in a patient with no known predisposing pathology. No prior reports of cases of solitary rectal ganglioneuroma have been published as far as is known, and the origin of similar lesions which have been reported at other sites in the gastrointestinal tract is a subject for speculation.

Ganglioneuromas are rare tumours and seldom encountered in the gastrointestinal tract. The first documented case was reported in 1928,¹ with subsequent reports predominantly concerning lesions of the stomach or duodenum, with a few reports of tumours in the large bowel.

Most ganglioneuromas arise in the posterior mediastinum, retroperitoneum, or adrenal in childhood or early adult life. Maturation of childhood neuroblastomas to ganglioneuromas has also been documented.²

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Figure 1 The upper border of rectal ganglioneuroma of case 1 showing single and clustered ganglion cells encroaching on the muscularis mucosa.

Although the gut is rarely affected, multiple diffuse ganglioneuromatosis has recognised associations. These include von Recklinghausen's disease, multiple endocrine neoplasia syndrome type 2B,³ Cowden's disease⁴ and juvenile polyposis.⁵ Most solitary lesions occur as isolated findings⁶ or as part of one of the above conditions. A search through published reports failed to uncover previous cases of solitary rectal ganglioneuromas.

Case reports

CASE 1

A 62 year old woman with a history of stubborn bowel habit was examined by sigmoidoscopy under general anaesthesia. The assessment was part of an annual surveillance after a total of six small colorectal tubulovillous adenomas had been excised on two occasions over the previous two years. At sigmoidoscopy, no abnormality was seen but for the presence of a 2.0 cm in diameter submucosal swelling in the posterior upper

Figure 2 Low power view illustrates the defined upper margin of case 2 abutting the mucosa. Ganglion cell clusters are indistinct at this magnification.

rectum, reported to be at a similar site to one of the previous polypectomies. The lesion was surgically removed.

CASE 2

A 79 year old man with a long history of indigestion, flatulence, and slight increase in stool frequency was referred to a general surgeon for investigation. Sigmoidoscopy was undertaken and showed a 1.5 cm in diameter submucosal lesion in the upper rectum. No further abnormalities were present and investigations were otherwise unremarkable. Biopsy of the rectal lesion was performed using a sigmoidoscope.

Neither patient had stigmata of von Recklinghausen's disease, Crohn's disease, or any other relevant medical or family history.

Histopathology

Each lesion was similar in appearance. The submucosa of the rectum had been infiltrated by a well circumscribed spindle cell tumour. This encroached on and fragmented the muscularis mucosa. Admixed with the randomly orientated swathes of wavy spindle cells were prominent ganglion cells, focally and in clusters (figs 1 and 2). Though seen throughout the tumours, ganglion cells were more prominent at the periphery. Nuclear pleomorphism and mitoses were not conspicuous. Chronic inflammatory cells and eosinophils were scattered through each lesion with a single lymphoid aggregate in case 1. This tumour also exhibited several small collections of foamy macrophages. The mucosa overlying each tumour appeared, histologically, to be unremarkable.

Immunohistochemical staining for S100 protein showed positivity in the spindle cell areas of each tumour, in satellite cells surrounding ganglion cell bodies, and in prominent nerve fibres associated with each lesion.

Discussion

The cases described depict solitary rectal ganglioneuromas, one arising at the site of excision of a tubulovillous adenoma. Ganglioneuromas have previously been described as isolated lesions in the stomach, small intestine, appendix and colon, but only as part of multiple ganglioneuromatosis in the rectum in one report.⁷

The stomach and small intestine are the most commonly affected sites for both solitary and multiple lesions. Only a single report of malignancy in a gastrointestinal ganglioneuroma has been recorded.⁸ This occurred in the small intestine of a patient with von Recklinghausen's disease.

The neoplastic nature of gastrointestinal ganglioneuromas has been challenged by several authors. A hamartomatous or "reactive" origin as a traumatic neuroma with accompanying ganglion cells has been postulated.⁹ Histologically similar lesions of

the duodenum, "neuromuscular and vascular hamartomas", and gangliocytic paragangliomas may be associated with many of the former lesions that are undoubtedly associated with mucosal damage such as that seen in Crohn's disease.¹⁰ Furthermore, the occurrence of areas of neuromuscular hyperplasia, which may include ganglion cells, is well recognised in Crohn's disease. The histology of the tumours presented shows well circumscribed lesions with no adjacent mucosal abnormality. Case 1 seems to have arisen in the 12 month period between sigmoidoscopic examinations and may be directly related to the site of previous surgery. Although a mucosal lesion may seem to have healed, the chronic inflammatory reaction within the tumour may be evidence of previous surgery.

The identification of a gastrointestinal ganglioneuroma necessitates the clinical and histological consideration of further lesions or associated pathology. Indeed, in several patients ganglioneuromatous lesions of the gut have been the presenting features of multiple endocrine neoplasia syndrome type 2b.

Addendum

Since the preparation of this paper the

occurrence of diffuse neuronal hyperplasia, in some cases including ganglion cells, has been highlighted in von Recklinghausen's disease by C E Fuller and G T Williams: Gastrointestinal manifestations of type 1 neurofibromatosis (von Recklinghausen's disease). *Histopathology* 1991;19:1-11.

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Rapid differentiation of *Mycobacterium xenopi* from mycobacteria of the *Mycobacterium avium-intracellulare* complex by pyrolysis mass spectrometry

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Abstract

Thirty four cultures of slow growing, Tween-80 negative mycobacteria were analysed by pyrolysis mass spectrometry. The results showed that pyrolysis mass spectrometry could positively distinguish strains of *Mycobacterium xenopi* from those of the *Mycobacterium avium-intracellulare* (MAI) complex. Pyrolysis mass spectrometry may be a useful technique for the rapid characterisation of non-tuberculous mycobacteria in such clinical settings as their isolation from immunocompromised patients—for example, those with AIDS.

Pyrolysis mass spectrometry is a rapid and simple technique for comparisons of strains of micro-organisms that has been successfully

applied to a wide range of bacterial species.¹ Pyrolysis mass spectra vary with the age and cultural conditions of the organisms before pyrolysis² so that pyrolysis mass spectrometry cannot assign permanent type designations. However, if organisms are prepared under identical conditions and then examined by pyrolysis mass spectrometry within a single machine batch, the system is highly discriminatory. Mycobacteria form a highly disparate group of organisms with a wide range of different growth cycles and optimal growth temperatures. Consequently, pyrolysis mass spectrometry is only likely to be useful when analysing within groups of mycobacteria of closely similar growth pattern. We recently adopted this approach in analysing members of the *M tuberculosis* complex. Pyrolysis mass spectrometry was able to distinguish *M tuberculosis* from *M bovis*, and also showed the close

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