Cap polyposis – an unusual cause of diarrhoea

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Abstract
‘Cap polyposis’ is a poorly recognised condition with distinct clinical, sigmoidoscopic, and pathological features that may be confused with other inflammatory conditions of the large intestine including pseudomembranous colitis and idiopathic chronic inflammatory bowel disease. The pathogenesis is unknown but on the basis of the characteristic histological appearances, which are similar to those seen in situations where mucosal prolapse is the underlying mechanism, it has been suggested that the latter may be an important aetiological factor. Two cases are described. Histological features in the first (presence of intramucosal elastin) and clinical features in the second (rectal prolapse) support the above hypothesis.

‘Cap polyposis’ was first described by Williams, Bussey, and Morson in 1985. This poorly recognised condition has distinctive clinical, sigmoidoscopic, and pathological features. The pathogenesis is unknown but the histological appearances resemble those seen in solitary rectal ulcer syndrome or in areas of prolapse – for example, adjacent to diverticula or in relation to prolapsing colostomies. It has therefore been postulated that prolapse may be an important aetiological factor. Two cases are described in which the clinical and histological features may point to a possible pathogenic mechanism.

Case 1
A 68 year old white man presented in September 1988 with weight loss (approximately 1 st (6 kg) over the previous year), lower abdominal pain, and persistent watery diarrhoea. He had not noticed any blood or mucus. Barium enema showed marked spasm in the mid sigmoid colon associated with a localised area of diverticulosis. This was not, however confirmed subsequently. Initial biopsy specimens were thought to be suggestive of pseudomembranous colitis but examination of stool for Clostridium difficile toxin was negative and treatment with metronidazole was unhelpful. Later sigmoidoscopy and biopsy showed changes thought to be more suggestive of inflammatory bowel disease with ‘pseudopolyps’. Treatment with sulphasalazine and prednisolone was unsuccessful and the patient underwent a left hemicolectomy in February 1990. The histological findings were multiple metaplastic polyps separated by normal mucosa with no evidence of inflammatory bowel disease.

Unfortunately the patient’s symptoms recurred and 5 months later he presented again with a 2 month history of persistent diarrhoea with weight loss (approximately 1 st (6 kg) over the previous year), lower abdominal pain, and persistent watery diarrhoea. He had not noticed any blood or mucus. Barium enema showed marked spasm in the mid sigmoid colon associated with a localised area of diverticulosis. This was not, however confirmed subsequently. Initial biopsy specimens were thought to be suggestive of pseudomembranous colitis but examination of stool for Clostridium difficile toxin was negative and treatment with metronidazole was unhelpful. Later sigmoidoscopy and biopsy showed changes thought to be more suggestive of inflammatory bowel disease with ‘pseudopolyps’. Treatment with sulphasalazine and prednisolone was unsuccessful and the patient underwent a left hemicolectomy in February 1990. The histological findings were multiple metaplastic polyps separated by normal mucosa with no evidence of inflammatory bowel disease.

Unfortunately the patient’s symptoms recurred and 5 months later he presented again with a 2 month history of persistent diarrhoea which now contained copious amounts of mucus. Sigmoidoscopy showed multiple haemorrhagic polyps in the rectum. These occurred on the apices of the transverse mucosal folds and were separated by normal mucosa. Microscopic examination of biopsy specimens showed ulcerated inflamed mucosa with elongated tortuous crypts attenuated towards the mucosal surface. Review of the sections taken from the previous colectomy specimen showed polyps at the apices of exaggerated mucosal folds with similar histological features to those described above. They were covered by a ‘cap’ of inflammatory granulation tissue (Fig 1). The intervening mucosa was normal. The appearances were those of cap polyposis.

The patient was treated with hydrocortisone and mesalazine enemas. These were unsuccessful and a total colectomy with ileostomy was performed, from which he made an uneventful recovery. Macroscopic examination showed sessile haemorrhagic polyps on the apices of mucosal folds over the distal 12 cm of the specimen. The mucosa between the polyps and proximal to them was normal (Fig 2). Microscopic features were as described above. Sections from the two resection specimens were examined with an orcein stain for elastin fibres. Intramucosal elastin was present in small quantities at the edges of some of the cap polyps (Fig 3).

Case 2
A 65 year old Belgian woman presented with a 6 month history of recurrent mucoid diarrhoea. Previous investigations had been inconclusive and a differential diagnosis of infection, solitary

Figure 1: Low power view of a typical polyp with granulation tissue cap (high power inset) (original magnification ×17; high power inset ×87)
rectal ulcer, or trauma had been made. She failed to respond to treatment with steroids and her symptoms had persisted after sigmoid colectomy with formation of a mucous fistula. On this occasion sigmoidoscopy showed evidence of rectal mucosal prolapse with 3-4 cm of perineal descent. The mucosa was oedematous and hyperaemic with distinct areas of intense inflammation on the mucosal folds and superficial ulceration. Biopsy specimens showed superficial erosion with elongated hyperplastic looking glands and a mixed inflammatory cell infiltrate. Review of the sigmoid colectomy showed prominent mucosal folds, vascular dilatation, and developing 'cap lesions'. An earlier biopsy showed the typical changes described above (Fig 4). The patient's symptoms have improved with claversal enemas and resection of the rectum is planned at a future date.

Discussion

Cap polyposis is a rare condition that may affect patients of both sexes and is seen over a wide age range (17-82 years). The commonest symptoms are mucous diarrhoea, tenesmus, and rectal bleeding. Haemorrhagic polyps separated by normal mucosa are seen in the rectosigmoid at sigmoidoscopy. These occur at the apices of the transverse mucosal folds. Histology shows that the polyps are composed of elongated tortuous crypts, attenuated towards the mucosal surface with a mixed inflammatory cell infiltrate in the lamina propria. The surface of the polyp is usually ulcerated and covered by a 'cap' of fibrinopurulent exudate and inflammatory granulation tissue. Treatment is by polypectomy or resection of the rectosigmoid.

Similar histological appearances are seen in other disorders where mucosal prolapse is the underlying mechanism – for example mucosal prolapse syndrome (solitary rectal ulcer syndrome and related disorders) and adjacent to large bowel tumours, diverticula, prolapsed colostomies, and haemorrhoids. It has therefore been postulated that prolapse may also be an important aetiological factor in the pathogenesis of cap polyposis. The coincident findings of overt rectal mucosal prolapse and cap polyps in our second case provide further support for this idea. Intramucosal elastin is not seen in normal large bowel mucosa but has been shown in solitary rectal ulcer syndrome and other conditions associated with mucosal prolapse. The presence of intramucosal elastin in small quantities at the edges of some of the cap polyps in the resection specimens from case 1 is an additional feature in favour of the above hypothesis.

Cap polyposis predominantly affects the rectosigmoid and may be seen in association with diverticular disease. It is possible that abnormal colonic motility, as seen in some patients with diverticulosis, may lead to prolapse of redundant mucosa at the apices of transverse mucosal folds. The resultant local ischaemia then produces the characteristic histological appearances. Cap polyposis, therefore, may be a variant of the
picture seen in crescentic colitis where inflammation characteristically affects the mucosal folds of the sigmoid colon in patients with diverticular disease.  

In conclusion, cap polyposis is a condition with distinct clinical, sigmoidoscopic, and pathological features that may cause confusion in the diagnosis of inflammatory bowel disease. Mucosal prolapse and abnormal rectosigmoid motility may be important in its pathogenesis.

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