LESSON OF THE MONTH

Dietary advice in systemic sclerosis: the dangers of a high fibre diet

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Systemic sclerosis frequently affects the gastrointestinal tract.¹ However, information on large bowel involvement and dietary advice for patients is scarce. Even the most recent and comprehensive rheumatological text discusses colonic involvement but gives no information on dietary advice or management.² It is widely felt and advertised that a high fibre diet is "good for all". We report on four cases, three of whom required emergency admission after receiving such advice.

Case reports

CASE 1

A 48 year old woman was admitted with a 10 day history of severe abdominal pain and distension. She had a four year history of diffuse systemic sclerosis and was taking penicillamine and indomethacin. Two weeks before admission she had recounted some difficulty with infrequent bowel motions to a friend, who had advised a high fibre diet. She obtained a high fibre breakfast cereal. Within three days she developed abdominal distension and colicky pain. This led to colonic atony with no bowel action for 10 days before admission. She was treated with daily picolax and discharged three days later. A barium enema did not reveal any obstructive lesion. She has remained well on her normal low residue diet and has had no similar problems or investigations since.

CASE 2

A 46 year old woman was admitted with a five day history of abdominal pain and vomiting. She had developed diffuse systemic sclerosis 10 years previously. Her symptoms had been well controlled with omeprazole and nifedipine. However, she reported to her doctor that she was having problems with constipation. He advised a high fibre diet with All-bran, wheat bread, and vegetables. Within a week she was admitted with a distended abdomen and vomiting. Plain films revealed a loaded colon. She was managed conservatively with picolax and discharged five days later on her usual low residue diet. She had no further problems or investigations until 1997 when she was admitted with abdominal pain, weight loss, and vomiting. Investigations confirmed small bowel overgrowth, which has responded well to cyclical antibiotic therapy.

CASE 3

A 71 year old woman with a nine year history of diffuse systemic sclerosis had noticed that she had an infrequent bowel habit. She was otherwise well and only took weekly methotrexate. She read in a women's magazine that a high fibre diet was a natural method of improving constipation. She started regular All-bran and increased her intake of fresh vegetables. Within a few days she developed increasing abdominal pain and distension. Recognising the change she immediately reverted to her low fibre diet. Her symptoms settled over a few days. She has remained well since and has required no further investigations.

CASE 4

A 56 year old woman developed systemic sclerosis aged 43. She was treated symptomatically but developed increasing problems with constipation. She consulted with her doctor who advised a high fibre diet and prescribed fybogel regularly. She was admitted shortly afterwards with acute abdominal pain and constipation. "Colonic atony" was diagnosed and a temporary colostomy was performed. This was then converted to a permanent ileostomy the following year. Since then she has had to stick rigidly to a low fibre diet. She remains underweight at 53 kg and has recurring problems with her stoma. Despite her considerable problems she continues to work full time.

Discussion

Systemic sclerosis is known to affect the whole of the gastrointestinal tract.¹ Histopathological studies have shown diffuse fibrosis of the colon with atrophy of the smooth muscle layers are a feature of this disease.³ As a result constipation or infrequent bowel movements are common complaints.^{4 5} Radiologically this may be seen as complete loss of colonic haustrations and dilatation similar to that seen in chronic ulcerative colitis.⁶ These changes are known to occur more commonly in the diffuse rather than the limited form of the disease. Resting colonic myoelectric activity is abnormal and responses to neostigmine and metoclopramide are blunted.⁷ In that study 9 of 10 of these patients had no colonic spike activity at all after eating. These pathological changes result in reduced colonic motility and prolonged transit

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times suggesting relative colonic atony.⁸⁻⁹ This may lead to a state of chronic colonic pseudo-obstruction.¹⁰⁻¹¹

Given the above it may be of little surprise to find that loading the bowel with dietary fibre acutely can lead to problems. There is some evidence to suggest that patients with systemic sclerosis have worked this out for themselves.¹² This dietary survey convincingly shows that fibre intake is significantly lower in patients with scleroderma when compared with healthy controls. Although these findings could also reflect that large meals may cause uncomfortable oesophageal reflux and so limit patients fibre intake, it seems probable that this is determined by patients previous experiences of "trial and error" as in one of our cases.

The precise mechanism of relative colonic atony in these patients is not as yet completely understood. Apart from the known fibrosis and myoelectric abnormalities, disturbances in the levels of gut regulatory peptides have been reported.¹³ Plasma concentrations of motilin, corticotropin releasing hormone, neuropeptide Y and peptide YY were all found to be significantly increased in scleroderma patients. It is not yet clear whether these are primary or secondary phenomena or how important these are in determining colonic motility.

While the colon in scleroderma is often hypomotile it is recognised that increasing stool bulk, and therefore colonic diameter, can improve colonic contraction. Thus the gradual introduction of fibre in the diet may soften stools and improve bowel symptoms in some cases. Alternatively osmotic agents such as lactulose may be also be tried with some success. Frequently an explanation of patients relative constipation while on a low fibre diet is acceptable and all that can be achieved.

All four patients we report here described considerable abdominal pain and three required hospital admission. The potential seriousness of this should not be underestimated, as both colonic perforation and even deaths have been reported in patients with scleroderma.¹⁴⁻¹⁶ In retrospect the use of picolax in two of these cases may be questioned, as this powerful colonic stimulant might increase the chances of perforation. Fortunately their pseudo-obstruction settled with these conservative measures without complication. Nevertheless, care should always be taken in such cases. Even a barium enema study should be carefully considered, as colonic

atony with barium impaction has been reported.¹⁷

The lessons

- If patients with systemic sclerosis present with symptoms of constipation, any advice to increase dietary fibre should be used with caution. Alternative methods including osmotic agents such as lactulose should be considered.
- If increased fibre intake is thought necessary it should be introduced gradually.
- In general these patients should avoid increased colonic loading and encouraged to accept infrequent bowel movements on a low fibre diet.

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