Ultrasonography to diagnose and exclude intussusception in Henoch-Schönlein purpura

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Abstract

Abdominal pain is a frequent symptom in the child with Henoch-Schönlein purpura and raises the suspicion of intussusception or perforation. One hundred and fifty two children with a diagnosis of Henoch-Schönlein purpura over 11 years were reviewed. Of these 60 had abdominal pain, 19 gastrointestinal bleeding, and nine were suspected intussusception. Intussusception was confirmed in two of these cases with ultrasonography.

Ultrasound is an important tool in the early diagnosis of intussusception complicating Henoch-Schönlein purpura. Where the intussusception appears loose an expectant policy, with careful monitoring, may allow spontaneous reduction. It may also be used in monitoring patients for postoperative recurrence of intussusception, mural haematoma, and uncomplicated intestinal vasculitis with oedema.

Abdominal pain and gastrointestinal bleeding are common manifestations of Henoch-Schönlein purpura, resulting from vasculitis, intestinal oedema, mural haemorrhage, intussusception, necrosis, and perforation.¹⁻³

Intussusception is an uncommon complication, yet its diagnosis is made more difficult in this situation.^{4 5} Not only is its clinical presentation less distinctive, especially when the child already had abdominal symptoms and signs, but definitive diagnosis and treatment by barium or air enema is made more hazardous in view of the underlying intestinal vasculitis and may not be accessible by the enema as it will usually be in the small intestine.⁶

Ultrasonography was useful in the diagnosis of intussusception in two recent cases. To try to establish the extent of the problem it was decided to review the surgical experience in all patients admitted to the hospital with Henoch-Schönlein purpura over an 11 year period.

Patients and methods

A chart review was made of patients admitted with the diagnosis of Henoch-Schönlein purpura from January 1980-December 1990 to determine the incidence of abdominal symptoms and surgical complications.

Results

Records of 152 patients were available for study, 60 of these experienced abdominal pain,

19 had gastrointestinal bleeding, and nine were suspected of having intussusception. Intussusception was excluded in most of the cases either by clinical examination, plain film of the abdomen, and more recently by ultrasound in five cases. It was confirmed in two cases by ultrasound, which constituted 1.3% of the patients analysed, and these are reported below.

CASE 1

A 5 year old boy was admitted in 1989 with a two day history of swelling of the feet and wrists, a purpuric rash on the ankles, scrotal erythema, vomiting, and intermittent abdominal pain. Urine analysis revealed proteinuria. A clinical diagnosis of Henoch-Schönlein purpura was made. His symptoms settled and he was discharged after six days.

One week later he was readmitted to the hospital with acute severe abdominal pain, tenderness all over the abdomen but no mass was palpable. A plain film of his abdomen was suggestive of intussusception, which was confirmed by ultrasound (fig 1). Hydrostatic or barometric reduction was not attempted in view of the underlying vasculitis. At laparotomy an ileoileal intussusception was identified and reduced manually with some difficulty. Though the bowel was congested looking it was viable with no perforation, and an appendectomy was done.

Initially he had an uncomplicated recovery,

Figure 1 Ultrasound scan of a large intussusception with typical 'swiss roll' appearance (black arrows) and surrounding free fluid (white arrow).

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but four days postoperatively he developed severe crampy abdominal pain. Abdominal ultrasound again demonstrated an intussusception in the right flank. At laparotomy on this occasion a caecocolic intussusception was reduced manually with little difficulty. A healthier looking small bowel was sutured side to side over a suspicious and slightly ischaemic looking patch at the site of the previous intussusception. The child recovered slowly and was discharged 18 days postoperatively.

Four days later he was readmitted with abdominal pain and bloodstained faeces. Abdominal ultrasound demonstrated thickened loops of bowel but no evidence of intussusception. He was treated conservatively. His renal involvement was treated with steroids. He eventually settled and was discharged home.

CASE 2

A 9 year old boy was admitted in 1990 with a seven day history of pain and swelling of his ankles and wrists, purpuric rash over both his legs and buttocks, and colicky abdominal pain. Two days after his admission, when his abdominal pain became more severe, an ultrasound was done which revealed an intussusception in the right flank, which the radiologist visualised to be occurring and reducing, and concluded to be 'loose' (fig 2). It was decided to observe him overnight with the expectation that it might reduce spontaneously, and to repeat the abdominal ultrasound in the morning. This revealed that the intussusception was no longer present. His abdominal symptoms improved.

Three days later he again started to complain of intermittent colicky abdominal pain. A plain film of his abdomen revealed absence of caecal gas shadow. The abdominal ultrasound confirmed an intussusception, which on this second occasion appeared tight. A laparotomy was carried out without delay and an ileoileal intussusception was easily reduced manually, with plication of the mesentery of the terminal ileum to prevent recurrence, and an appendectomy was

done. He gradually improved and on recovery was discharged home.

Discussion

Some authors have drawn attention to the difficulty in the diagnosis of intussusception in Henoch-Schönlein purpura.^{4 5 7} Identification of patients with this complication may be obscured for a number of reasons:

(1) Abdominal symptoms are frequent among patients with Henoch-Schönlein purpura who do not have intussusception.

(2) The presentation of intussusception is variable and may be identical to that of intestinal vasculitis and oedema, or mural haematoma, not requiring surgery.^{2 3 5}

(3) Intussusception may occur at any stage of the illness.^{2 5}

(4) After surgery intussusception may occur at a different site as seen in our first case.

(5) The place of contrast studies is limited by its invasiveness, and by the fact that most intussusceptions in Henoch-Schönlein purpura are in the small bowel.⁶ Especially in the presence of vasculitis with severe intestinal involvement, there may be a high risk of perforation.

A previous report has described the use of ultrasound in the diagnosis of a case of intussusception in Henoch-Schönlein purpura.⁶ Our report highlights the use of ultrasound to diagnose or exclude intussusception as a complication of Henoch-Schönlein purpura. It is also useful as an efficient, non-invasive, and repeatable method in the postoperative period to identify or exclude recurrence or further intussusception. It may provide a way of carefully monitoring cases in which the intussusception is loose and may reduce spontaneously.

Our study confirms that abdominal symptoms are present in a significant number of children with Henoch-Schönlein purpura. Perforation of bowel is easily diagnosed on a plain film of the abdomen and was not seen in our group of patients. Intussusception was seen, but the incidence was lower than that reported in other series.^{1 3}

A total of 60 patients spread fairly evenly throughout the 11 year review had abdominal pain, but six of the nine suspected cases of intussusception occurred in the last two years. It is possible that intussusception in Henoch-Schönlein purpura is a more common occurrence than we have found, and that the intussusception may be loose and may reduce spontaneously as was shown by ultrasonography in our case 2. When a second intussusception occurred in this case, it was described as tight and therefore laparotomy was carried out immediately.

The intussusception was reduced easily and it is possible that it would have reduced had an expectant policy been followed. Despite this, in our current state of knowledge, we would have to recommend that the surgical line be followed when the ultrasound picture is suggestive of a tight intussusception. However, it would seem reasonable to follow a 'wait and observe' policy if it appears loose with serial ultrasonography.

We would not consider attempting a therapeutic air enema reduction (or any other con-

Figure 2 Small loose intussusception (arrows) which reduced spontaneously.

trast enema) as the bowel is generally inflamed, haemorrhagic, and oedematous as a result of the vasculitis, making perforation a significant risk, and also as the intussusception is usually in the small bowel and will not be accessible by this form of treatment.⁶

Patients in whom laparotomy has been avoided by the use of ultrasound can also be usefully monitored by serial ultrasound examination to assess the resolution of free fluid, intestinal oedema, and mural haematoma. Careful clinical monitoring is essential in patients with Henoch-Schönlein purpura with prominent abdominal symptoms, complemented when necessary by serial ultrasound examinations. Not only can the clinician thus identify the complication of intussusception at an early stage, but also needless laparotomy may be avoided in

patients with uncomplicated intestinal vasculitis and oedema.

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Enzyme replacement in Gaucher's disease

Gaucher's disease is due to a deficiency of the enzyme glucocerebrosidase in macrophage lysosomes. Giving placental glucocerebrosidase intravenously gave poor results probably because the enzyme was taken up by hepatocytes which do not accumulate glucocerebroside in this disease. For more success, so the theory goes, the enzyme must be delivered to the affected macrophages. That can be done by taking advantage of the special properties of macrophage plasma membranes. American workers have prepared macrophage-targeted glucocerebrosidase by sequential deglycosylation of the oligosaccharide chains of the enzyme and a recent report in the *New England Journal of Medicine* (Norman W Barton and colleagues, 1991;324:1464-70) describes the effects of its administration to eight children and four adults with non-neuronopathic Gaucher's disease.

The enzyme was given by intravenous infusion every two weeks for nine to 12 months. There were no apparent adverse effects. A significant reduction in spleen size occurred in all 12 patients with an accompanying rise in haemoglobin concentration. Serum acid phosphatase activity fell in 10 patients and liver volume in five. Plasma glucocerebroside concentrations fell in nine patients. The changes occurred slowly over six to nine months or more. Bone pain improved and three patients showed improvement in long bone x ray changes though the skeletal effects were even slower than the visceral or haematological. All the patients reported increased wellbeing, more enthusiasm for daily activities, and reduced fatigue.

Clearly these results are extremely encouraging. The authors comment that the same approach may benefit people with other metabolic diseases. Is this the dawn of a new era of targeted enzyme therapy or is it a false dawn? Keep your fingers crossed.

ARCHIVIST