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Acute Gastrointestinal and Genito-urinary Manifestations in Children with Henoch-Schönlein Purpura

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Abstract

Purpose: To review the incidence and management of acute gastrointestinal and genito-urinary manifestations in children with Henoch-Schönlein purpura (HSP). ***Methods:*** The records of children admitted with HSP in recent 10 years were reviewed. ***Results:*** 165 children were admitted with HSP. Seventy children, aged 2-17 years, had acute surgical manifestations, 42 of which presented before the onset of purpura or arthralgia. Gastrointestinal symptoms included abdominal pain (61), per-rectal bleeding (9), prolonged ileus with ascites (1) and haematemasis (1). Genitourinary symptoms included acute scrotum (9) and frank haematuria (2). Abdominal sonography was performed in 14 patients, scrotal sonography in two boys. None required operative intervention. Twenty-seven children required systemic steroid. ***Conclusions:*** Acute gastrointestinal and genito-urinary manifestations are common in children with HSP and frequently precede purpura and arthralgia. Vigilant physical examination and judicious sonography enable correct diagnosis. Conservative management almost invariably suffices and systemic steroid is useful in selected cases.

Key words Gastrointestinal; Genito-urinary; Henoch-Schönlein purpura; Surgical manifestations

Introduction

Henoch-Schönlein purpura (HSP) is a disorder of systemic small vessel vasculitis of unknown aetiology. It affects children with a peak incidence between 3 and 8 years of age. Schönlein, in 1837, first described the characteristic purpura and arthralgia, which are together pathognomonic of HSP. Some forty years later Henoch recognised the frequent association with gastrointestinal and genitourinary symptoms in this condition.

Abdominal manifestations, which may occur in up to two-thirds of all children with HSP, are due to gastrointestinal tract involvement with submucosal and subserosal oedema and haemorrhage, or complicating nephritis. Symptoms may vary from non-specific pain and vomiting with ileus and ascites, to massive upper or lower gastroinestinal haemorrhage and frank haematuria. Acute scrotum has also been described in up to 25% of boys with HSP. These conditions often present acutely to both physicians and surgeons, but are by and large self-limiting and seldom require surgical intervention if diagnosed correctly.

Other specific HSP associations such as intussusception or bowel perforations, on the other hand, are rare but have life-threatening consequences if missed or untreated surgically.

In this study we review the incidence and the

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management of these surgical manifestations of HSP in the paediatric population of our locality, and compare with those reported in the literature elsewhere.

Methods

In two cluster-hospitals, Queen Elizabeth Hospital (Kowloon Central) and United Christian Hospital (Kowloon East), we searched the records of all the children admitted with the diagnosis of HSP in a recent ten year period. We identified those with surgical manifestations, i.e. significant abdominal pain, gastrointestinal upset, frank haematuria, acute scrotum, and other specific causes for surgical or acute abdomen. By collecting patient data using a standardised proforma, we reviewed the incidence and timing of these symptoms, the management in terms of diagnostic radiology, surgical intervention if any, and their outcomes in terms of symptom duration and the need for systemic steroid.

Results

Between the two cluster-hospitals over a ten year period a total of 165 children were diagnosed clinically with HSP, all having symptoms of purpura with joint and/or abdominal manifestations. Seventy children (42%) had significant surgical manifestations documented in their hospital records. They were aged between 2 and 17 years (mean 6.6 years), with boy to girl ratio of 1.2 to 1.

The incidence of each surgical manifestation and pathology is listed in Table 1. In 42 children (25% of all HSP presentations), abdominal symptoms preceded the onset of purpura or arthralgia. In 26 of them, purpura, arthralgia and the diagnosis of HSP were not apparent at initial presentation to hospital clinician.

Table 1 Incidence of surgical manifestations in HSP

Surgical manifestations	Incidence (n=165)
Nonspecific abdo pain/vomiting	61
Lower GI haemorrhage	9
Acute scrotum	9
Frank haematuria	2
Ascites with ileus	1
Upper GI haemorrhage	1
Surgical abdomen (e.g. intussusception/perforation)	0

GI: gastrointestinal

Concerning radiological investigations, 14 children with significant signs or symptoms had abdominal sonography, none yielding specific surgical pathology. Four patients proceeded to have abdominal computer tomogram (CT) for severity or prolonged duration of symptom, but all were normal. Nine boys presented with acute scrotum, five of whom had unilateral involvement. Two of them were suspected of testicular torsion and had scrotal sonography and doppler study; they were both positive for testicular blood flow.

Symptom durations vary from less than one day to 60 days. Twenty-seven children had systemic steroid administration for more severe symptoms, with the mean duration of symptoms in this group being 7.3 days. Among those who did not require steroid therapy the mean symptom duration was 3.4 days. The overall length of hospital stay varied from one day to 28 days, with a mean of 10.6 days.

None of these children with gastrointestinal or genitourinary HSP manifestations were found to have any surgical causes requiring definitive treatment or operation.

Discussion

The surgical manifestations of HSP were frequent in our locality, a great majority being non-specific gastrointestinal upset and abdominal pains, which resolved with conservative treatment. In fact the quoted incidence of abdominal symptoms in studies elsewhere has been even higher. Katz et al reported more than 65% gastrointestinal involvement in a review of 110 cases.[1,2] Lower gastrointestinal bleed was the next most common, but were self-limiting in all our cases. None were severe enough to require endoscopic treatment or blood transfusion.

Many other specific abdominal emergencies have been described in association with HSP, the most common being intussusception, occurring in up to 3.6% of cases.[3] Acute appendicitis, mimicked by vasculitic involvement of the appendix is reported to be misdiagnosed in up to 5% of children with HSP, resulting in unnecessary appendicectomies. In this series we encountered neither of these two conditions. Other reported rarities include bowel ischaemia and infarction (in the absence of

intussusception), spontaneous perforation of small bowel, massive intestinal haemorrhage, small bowel fistulae, pseudomembranous colitis, acute pancreatitis, gall-bladder involvement, and late ileal strictures.[3-9] Some of these require urgent surgical attention and definitive operative treatment to prevent life-threatening sequelae.

HSP associated nephritis is a result of vasculitic involvement of the kidneys causing haematuria, which is by and large microscopic type. Frank haematuria is uncommon in this series and is self-limiting in each case. In fact, in one patient it may have been a result of iatrogenic injury after a closed renal biopsy for nephritis workup, rather than due to the nephritis *per se*.

Acute scrotal vasculitic involvement of the scrotum results in hydrocele, oedema of scrotal soft tissue, testis and epididymis which mimic torsion and other causes of acute scrotum. Reported incidence is up to 24% in boys with HSP, with up to 60% of these being unilateral.[10] In our series only nine boys (10%) were noted to have acute scrotum at presentation. Two of the five with unilateral involvement required ultrasound evaluation for testicular flow. This compares favorably with audit by Ioannides et al in which the scrotal sonography rate was 14% and the surgical exploration rate was 36%.[11] The underlying diagnosis of HSP should, in most cases, obviate any need for surgical scrotal exploration. The acute scrotums invariably resolve with conservative treatment, but in cases of doubt with unilateral involvement, sonography and doppler study should demonstrate the presence of arterial flow as well as typical inflammatory changes associated with HSP, avoiding unnecessary operation.

When abdominal manifestations become severe or prolonged, specific surgical complications must be considered. Laboratory tests tend to be unhelpful in differentiating surgical causes. Serial physical examinations and appropriate radiological work-up must follow. Early plain radiographs often demonstrate non-specific dilated bowel loops and thickened bowel walls. Evidence of distal ileal obstruction with 'cut-off', as in intussusception, and perforation must be excluded. Cull et al recommended sequential barium enema and upper contrast series to exclude intussusception.[1,2] This is now replaced in many centres by sonography. It is non-invasive and free of radiation, making serial assessment more acceptable to the child. Although operator dependent, sonography can accurately demonstrate mural thickness, mural haematoma, ileus, peritoneal fluid and intussusception.[12] The role of abdominal sonography has been well documented in diagnosing intussusception and is the investigation of choice in these clinical circumstances.[13,14] Computerised tomography is now an accessible and liberal diagnostic tool; but in our series it provided little information additional to sonography and served merely as a helpful reassurance in excluding surgical pathology.

Acute abdominal signs and symptoms may potentially be masked by early systemic steroid therapy. Judicious use of imaging in suspicious cases is important. When life-threatening surgical complications are excluded after serial physical examinations and radiological workup, the physician may step up systemic steroid therapy for symptom alleviation.

In 25% of cases in this HSP series surgical signs and symptoms preceded the pathognomonic rash and arthralgia. When assessing these patients, the clinician must be vigilant throughout to avoid misdiagnosis and inappropriate surgical treatment, since conservative measures, with or without steroid therapy, suffice in a great majority of them. Conversely for children with known HSP, the clinician must also be vigilant for any life-threatening complications that very occasionally develop. Serial surgeon's assessment and judicious sonography will prevent delay of timely definitive surgical management.

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