Dear Editor,

LEFT HAND COMPARTMENT SYNDROME: A RARE COMPLICATION OF HENOCH SCHONLEIN PURPURA

Henoch Schonlein Purpura is the most common vasculitic disease of childhood.\(^1\) It is a multisystem disease most commonly affecting skin, joints, gastrointestinal tract and kidneys. Other rare manifestations include seizures, scrotal swelling, acute cholecystitis and myocardial infarction. However, acute isolated limb compartment syndrome has not been reported. We report a case of a boy with Henoch Schonlein Purpura complicated with isolated left hand compartment syndrome.

A 9-year old boy was admitted for severe left iliac fossa pain associated with an episode of haematochezia. There was a preceding history of purpuric rash over his lower extremities associated with arthralgia 2 weeks earlier.

On examination he appeared well with a blood pressure of 110/60 mmHg (90th centile). Purpuric lesions were seen over both legs. Abdominal examination revealed generalised tenderness without guarding. Ultrasound of his abdomen confirmed no evidence of intussusception. During hospitalisation, he had a transient left scrotal swelling which resolved spontaneously. Due to his persistent abdominal pain, he was started on intravenous Hydrocortisone 2 mg/kg/day for 3 days and was discharged 3 days later with oral Prednisolone 1 mg/kg/day.

He returned 4 days after discharge with severe abdominal pain. Repeated ultrasound abdomen showed no evidence of intussusception. However, he developed fresh per-rectal bleed that required blood transfusion and intensive care monitoring.

His left radial artery was cannulated for a period of 36 h for invasive blood pressure monitoring. About 20 h after removal of the arterial line, the entire left hand became swollen and tender with extensive purpuric lesion up to the midforearm. He was referred to the Orthopaedic Hand Surgery Team and a diagnosis of Left Hand Compartment Syndrome was made. Doppler flow imaging showed no evidence of venous or arterial thrombosis. An emergency Left Hand Fasciotomy Decompression was done which revealed findings of transudative fluid in the thenar compartment, second and third dorsal compartment as well as haematomas in the hypothenar compartment.

Laboratory investigations showed normal haemoglobin level of 14 g/dL, normal total white cell count of 8.3 \(\times 10^9/L\) and normal platelet count of 250 \(\times 10^9/L\) as well as normal coagulation profile. Urinalysis and renal function test were normal. His C-reactive protein was low at 0.6 mg/L. His autoimmune markers were essentially normal, that is, normal C3 and C4, normal anti ds-DNA with a weak positive anti-nuclear factor (1:40) nucleolar pattern (which can be present in 20–30% of normal individual without any clinical evidence of connective tissue disease).

He was treated with IV Methylprednisolone 1 mg/kg/day for 10 days and was discharged well with tapering dose of oral prednisolone for 2 weeks.

Postoperatively, his wound healed well and no secondary suturing was required.

Henoch Schonlein Purpura is an inflammatory disorder of unknown cause characterised by IgA- dominant immune complexes in smaller venules, capillaries and arterioles.\(^2\) Its major complications include renal and gastrointestinal involvement. Our patient experienced purpura, abdominal pain with bloody diarrhoea, arthralgia, scrotal swelling and later complicated by an isolated left hand compartment syndrome.

Hand compartment syndrome as a complication of Henoch Schonlein Purpura is a very rare entity. To our knowledge this case report might be the first case reported in the literature. Whether spontaneous bleeding as a result of the vasculitic lesion of Henoch Schonlein Purpura is the cause of the compartment syndrome or whether it was due to the left radial artery cannulation that might have triggered a thrombus formation is difficult to ascertain.

However, the Doppler flow imaging showed no thrombus formation within the left hand vascular system. Furthermore, intraoperatively, haematomas were found in few areas and this finding is more suggestive of spontaneous vasculitic bleed. In this case, spontaneous vascular bleed as a result of Henoch Schonlein vasculitis may have caused the compartment syndrome. Hence we propose that compartment syndrome is a rare feature of Henoch Schonlein vasculitis.

References


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Dear Sir,

PACHYONYCHIA CONGENITA

Pachyonychia congenita is a rare genodermatosis which is autosomal dominant in inheritance. We describe a Malay toddler who presented with leukaemia and this condition.

A 20-month-old Malay boy was referred to our hospital for the management of acute lymphoblastic leukaemia. He had been noted to have abnormal nails at infancy and had been initially diagnosed to have onychomycosis. His nail clippings grew Aspergillus niger and he had been treated with itraconazole for several months. However, there was no improvement in the condition of his nails. He was then labelled as having yellow-nail syndrome.

On examination at our centre, he was found to have hyperpigmentation and hyperkeratosis of all his fingernails as well as toenails. His mother had similar findings (Fig. 1) and she also had hyperkeratosis of her soles. No one else in the family was affected. Apart from the nail changes, the child did not have any oral lesions and his palms and soles were also normal. A diagnosis of pachyonychia congenita type 1 was made clinically and his mother was counselled regarding the disease. The child has completed two years of chemotherapy without complications and is currently well and in remission.

Classic type 1 pachyonychia congenita (Jadassohn–Lewandowski syndrome) is due to mutations in the gene for keratin 16. It can cause a variety of ectodermal defects affecting the nails, skin, oral mucosa, larynx, hair and teeth. Nail dystrophy is the most striking feature, with thickened, tubular nails which can present either at birth or early in life as in our patient. The second most prominent feature is palmar-plantar keratoderma, which was present in this child’s mother. Other features include onychogryphosis, follicular keratosis and oral leukoplakia. Type 2 pachyonychia congenita is characterised by natal teeth and the development of epidermal cysts or seatoectysts.

Onychomycosis is also a cause of hyperkeratosis. However, it rarely affects all 20 nails. The most common cause of tinea unguium is the dermatophyte Trichophyton rubrum. In Malaysia, it has been found that moulds, in particular Aspergillus niger, cause a significant proportion (35%) of onychomycosis. The yellow nail syndrome is a triad of thickened, excessively curved, slow-growing yellow nails, primary lymphoedema and chronic respiratory disease. It has previously been classified as a dominantly inherited condition with variable expressivity. However, this has been challenged recently by findings that suggest that the majority of cases are sporadic and that the nail changes may remit. Our patient had no previous medical history of note; he had never had lymphoedema or respiratory problems.

The yellow nail syndrome is known to be associated with malignancies, in particular Non-Hodgkin’s lymphomas and carcinomas. In contrast, pachyonychia congenita has not been associated with cancers although squamous cell carcinoma has been reported in a patient who had chronic plantar ulcerations. It is likely that the leukaemia in our patient was a chance association.

The treatment of pachyonychia congenita consists of keratolytic agents and lubricants for the palmar and plantar hyperkeratosis. At present, there is no satisfactory treatment for the nail changes.

References

Dear Editor,

ON-LINE HEALTH INFORMATION FROM A CHILDREN’S HOSPITAL: USER FEEDBACK

Second to consulting a health-care professional, the Internet is one of the most common sources of health information for parents.1–4 Eighteen per cent of parents in an Australian study indicated that information they read online altered their health-care decisions3 – yet many health web sites have been found to be incomplete, inaccurate or not written at an appropriate reading level.5–7

In 2004, a group of Royal Children’s Hospital staff launched a collection of parent information fact sheets online for public access (http://www.rch.org.au/kidsinfo). These factsheets, covering acute and chronic complaints and diseases and in-hospital tests and procedures, now number in excess of 200 and receive more than 1500 visitors a day. The goal was to provide evidence-based and medically accurate information, presented in an appropriate and accessible way. While factsheets were developed in response to requests from parents and health-care providers, it was not clear if they were actually meeting the needs of the web site’s target group.

To facilitate ongoing improvement we analysed the voluntary online survey attached to each factsheet between September 2006 and May 2007. The survey was approved as an audit by the hospital Ethics Committee. The survey was composed of two parts. Part A asked about the usefulness of the factsheet. Participants could then choose to complete Part B, containing four more questions and an area for any comments.

During the 8-month study period there were approximately 360 000 visitors to the Kids Health Info web site. There were 574 survey responses to Part A and 308 to Part B representing 0.16% and 0.06% of visitors respectively. Web site usage statistics indicated that 63% of all visitors found it through a google.com or google.com.au search.

The 574 respondents to Part A had accessed 142 different factsheets; 63% of respondents thought the factsheet they read was ‘very useful’, 24% ‘useful’, 4% ‘not useful’ and 1% thought it was ‘really bad’. Seven per cent gave a ‘neutral’ rating.

The data from the 308 respondents to Part B indicated that 66% were viewing the factsheet for their own child and 16% were health-care providers accessing the factsheets for a patient. The three most common reasons respondents accessed the factsheets were ‘to find out usual treatment’ (54%), ‘causes’ (52%) and ‘how to help a child at home’ (42%) (Fig. 1). Ninety-three per cent rated the information as ‘easy to understand’; 83% said the factsheet they wanted was ‘easy to find’ and 81% thought the factsheet ‘answered all their questions’. Twenty-four per cent thought the factsheet needed more detailed information.

Sixty-three respondents provided constructive feedback that could be used to review or update factsheets either immediately or during bi-annual reviews. For example, evidence-based treatment options were updated following paediatrician feedback, information on the impact of certain diseases on families was altered based on feedback of families and information about local support agencies was added. Wherever possible, parents were involved in the review process.

In contrast to our expectations that respondents would mainly complete the survey if they were dissatisfied, most online survey feedback for the Kids Health Info web site was positive. Feedback indicated that overall, factsheets were written appropriately and clearly, were of the right length and were meeting a need. Although there is limited comparable literature, an NHS study of parent feedback on immunisation information indicated that 20% were not satisfied, mainly because the information was insufficient.8

A small but important number of factsheets were identified that needed significant review. Constructive suggestions could be addressed either immediately or during the next update, depending on the urgency of the recommendation.

Parents mainly used the web site to find out about usual treatment, causes and care at home for their child’s problem.

Fig. 1  Online health information – user feedback.
This information was used to review the standard template for developing new factsheets. Of interest is that 12% of respondents were accessing the factsheet to find out who they should see for medical attention. This suggests that the factsheets may influence health-care seeking behaviour among parents.

It is unclear how representative this sample is compared to the large number of visitors (360 000) over the study period. Survey feedback has also been limited to an English-speaking, Internet-using audience only.

In summary, the online feedback survey for a child health information web site indicated that overall, factsheets were written appropriately and meeting a need. To become and remain a valued and trusted resource for parent information on child health issues, consumers can and should play an important part of an ongoing improvement process.

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References


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