CASE REPORT

Unexplained bruising: a developing story

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SUMMARY

We present the case of a 10-month-old male infant who presented to accident and emergency with unexplained bruising of the ear. Initial blood tests showed no clotting or platelet abnormalities and non-accidental injury investigation commenced. He was subsequently reviewed by the dermatology team who suggested the diagnosis of acute haemorrhagic oedema of infancy presumably triggered by a viral or bacterial infection, and the clinical findings fit this diagnosis.

BACKGROUND

Acute haemorrhagic oedema of infancy (AHOI) is a rare cutaneous vasculitis which can appear very striking but is self-limiting and benign in nature. By highlighting this condition and its presentation, we hope to aid diagnosis of this unusual cause of purpura in children, possibly avoiding unnecessary diagnostics and procedures. In addition, it emphasises the importance of considering rarer diagnoses when non-accidental injury is one of the differential diagnoses in children presenting with unusual or unexplained signs.

CASE PRESENTATION

A 10-month-old boy was brought to paediatric accident and emergency by his mother with a 1-day history of unexplained purple discolouration affecting his right ear. On presentation, he appeared well, and physical examination revealed purpura on his right pinna with surrounding petechiae (figure 1). He also had petechial spots on his left pinna, right upper arm and buttocks. There were bilateral erythema and desquamation of the axillae, erythema and swelling below the penis and swelling of the right ankle. He had significant bilateral cervical lymphadenopathy. The infant was not feverish initially but spiked a fever of 39.2°C while in the department. He had a history of coryzal symptoms for 1 week and had been diagnosed with upper respiratory tract infection the previous day by his general practitioner, at which point none of the above signs were present. There was no history of trauma and no family history of clotting disorders. He is the youngest of five children, the oldest being 6 years old. There was no history of social services involvement. Over the following 24 hours, he developed a worsening purpuric rash on his buttocks spanning the natal cleft (figure 2), three bruise-like lesions on the sole of the feet and purpura over the right ankle (figure 3). All the purpuric areas were associated with mild oedema. Aside from the temperature spike to 39.2°C, he had

no fever for the duration of his admission so the evolution of the rash was not related to fever.

INVESTIGATIONS

Full blood count revealed raised white cell count of $25.6 \times 10^9/L$ and neutrophilia.

Platelets were raised at $528 \times 10^9/L$. C-reactive protein was 8 mg/L, and clotting screen was normal. Respiratory panel was sent off to try to identify the trigger for AHOI as it can be caused by viral or bacterial infections. Nasal swab showed scanty growth of *Staphylococcus aureus* sensitive to flucloxacillin, and nasopharyngeal aspirate was positive for rhinovirus and metapneumovirus. Chest X-ray and ankle X-ray, to investigate the bruising and swelling, were also reported as normal with no evidence of bony injury.

DIFFERENTIAL DIAGNOSIS

A diagnosis of non-accidental injury was initially considered given the presentation of an infant with unexplained bruising along with an unusual pattern of petechiae and joint swelling; therefore, child safeguarding procedures were initiated. We were unsure of a unifying cause for the purpura, petechiae, swelling and skin desquamation so a dermatology opinion was sought. The appearance of the

Figure 1 Bruising and petechiae of right ear on day of presentation to accident and emergency.

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Figure 2 Bruising over buttocks spreading on day 2 of admission.

skin under the axillae led to consideration of staphylococcal scalded skin. The possibility of non-accidental injury was disregarded after dermatology confirmed a diagnosis of AHOI, and social services had also conducted an initial assessment which raised no concerns. Table 1 details other differential diagnoses to be considered when assessing a patient with bruising and petechiae.

TREATMENT

The infant was started on intravenous flucloxacillin for suspected staphylococcal skin infection in view of the erythema and desquamation of both axillae. He was treated for 48 hours with intravenous antibiotics and then completed a 7-day oral course. He was observed for a further 24 hours and noted to have further areas of purpura/bruising developing during his stay which helped support the diagnosis. There is no specific investigation or treatment for this condition, other than treating the underlying triggering infection.

OUTCOME AND FOLLOW-UP

The purpura affecting the ear, buttocks and ankles were resolving by discharge. The oedema of the penis also improved but he remained coryzal. He was followed up in clinic in 2 weeks to ensure resolution of symptoms.

DISCUSSION

Snow first described this condition in the early 1900s,¹ and it has since been documented in the literature under various terms (Finkelstein disease, Seidlmayer syndrome, infantile postinfectious iris-like purpura).

Figure 3 Initial swelling of the right ankle and bruising that developed on day 2 of admission.

Fiore *et al* conducted a large systematic review of existing case reports in 2008, which included 294 children in total.² In these cases, the ages of children affected ranged from 2 to 60 months, with a male predominance (67%). The majority of cases had a history of prodromal illness, most commonly respiratory tract or diarrhoeal infections, with a small minority presenting following vaccinations. As with our case, the purpuric plaques were mainly seen affecting the extremities, cheeks and ears, with relative sparing of the trunk. Commonly, children present with low-grade fever (45%) but otherwise appear quite well.²

Initially AHOI was thought to be a mild variant of Henoch-Schönlein purpura (HSP), with the differences in presentation being due to age. However, current evidence suggests that it is a separate disease process with several differences in both presenting features and biopsy findings.²⁻⁴ As mentioned above, the age group affected tends to be younger than that of HSP and is generally limited to cutaneous signs and symptoms. Involvement of other systems is found in fewer than

Table 1 Differential diagnosis petechial rash⁹

Infection	
Meningococcal sepsis	Non-accidental injury
Enterovirus	Leukaemia
<i>Haemophilus influenzae</i>	Henoch-Schönlein purpura
	Thrombocytopaenia (eg, Immune thrombocytopaenia)
	Haemolytic-uraemic syndrome
	Clotting factor deficiencies

10% of cases reported.^{2,4} In cases where skin biopsies have been performed, histopathology shows a leucocytoclastic vasculitis similar to HSP. However, immunohistological investigations suggest that IgA deposition is rarely found with AHOI (<25%),² and smaller studies have suggested differences in activation of the complement system with C1q deposition present in AHOI but not HSP.⁴

The differences between the two conditions can be highlighted through the European League Against Rheumatism/Paediatric Rheumatology International Trials Organisation/Paediatric Rheumatology European Society diagnostic criteria for HSP. This requires palpable purpura as well as at least one of the following: diffuse abdominal pain, arthritis or arthralgias, renal involvement or leucocytoclastic vasculitis with predominantly IgA deposition,⁵ all of which are uncommon with AHOI.²

In terms of prognosis, true recurrence of AHOI after remission has not been reported,² which is also in contrast to HSP. Though generally self-limiting with few complications, note is made of specific cases in the literature, including one where an infant developed compartment syndrome as a result of oedema,⁶

and one case of intussusception.⁷ In addition, haemorrhagic lacrimation and epistaxis have been reported as rare complications of AHOI.⁸

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Learning points

- Acute haemorrhagic oedema of infancy should only be diagnosed once other potentially severe conditions such as meningococcal disease, Henoch-Schönlein purpura and non-accidental injury have been ruled out.
- Once other causes of purpura have been excluded, family and clinicians can be reassured that despite its alarming presentation, this condition is usually self-limiting with a good outcome.
- No treatment is recommended unless indicated for the underlying triggering infection.

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