Acute Haemorrhagic Oedema of Infancy (AHOI): A Case Report

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Abstract Acute haemorrhagic oedema of infancy (AHOI) is a benign variant of leukocytoclastic vasculitis which occurs in children up to 2 years of age. It is considered by some to be a variant of Henoch–Schönlein purpura with its hallmark of prominent facial swelling, purpuric rash without visceral involvement, in an otherwise well child. This condition is well recognised in the paediatric and dermatology literature but despite its impressive facial features, often mimicking more serious pathology like orbital cellulitis, to our knowledge AHOI has not been published in the Oral & Maxillofacial Surgery literature. We present a case of AHOI to raise awareness of this condition in maxillofacial surgery to avoid it being mis- or over-diagnosed.

Keywords Facial · Oedema · Acute · Haemorrhagic

Introduction

Acute haemorrhagic oedema of infancy (AHOI) is a benign variant of leukocytoclastic vasculitis which occurs in children up to 2 years of age [1–3]. It is considered by some to be a variant of Henoch–Schönlein purpura [2–4] with its hallmark of prominent facial swelling, purpuric rash without visceral involvement, in an otherwise well child. This disorder is recognised in the Paediatric and Dermatology literature, but has to our knowledge, not been

previously documented in the Oral & Maxillofacial Surgery literature despite its impressive facial features.

Clinical Case

A normally healthy, 2 year old female, was referred to the maxillofacial surgery team with facial and periorbital swelling. There was a 3 day history of fever, anorexia, generalised purpuric rash and fleeting, non-pitting oedema of the limbs and face.

On examination the child was alert and interactive. There was a generalised purpuric rash and swelling over the limbs (Figs. 1, 4), along with a non-erythematous, non-pitting, diffuse swelling of the right periorbital, frontal and temporal regions (Fig. 2). The right eye was closed but there was no obvious visual compromise. Erythema and swelling of the right ear was also noted (Fig. 3). Investigations showed an isolated raised CRP of 160 mg/l.

A provisional diagnosis of AHOI was made and supportive treatment with intravenous fluids and Ceftriaxone was commenced. The right periorbital oedema resolved in 24 hours but was followed by a similar swelling of the left periorbital tissues. The patient made an uneventful recovery over the following few days.

Discussion

AHOI, typically presents in children under 2 years of age with a recent history of upper respiratory tract infection, otitis media or recent vaccination. A generalized purpuric rash is present over the limbs with truncal sparing. The associated, impressive facial swelling is usually non-erythematous, non-pitting and can affect the face, ears, eyelids,

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Fig. 1 Widespread purpuric rash over lower limbs

Fig. 3 Erythema and swelling of right ear

Fig. 2 Periorbital oedema

cheeks and mucous membranes. Although there may be an associated low grade pyrexia $<38.5^{\circ}\text{C}$, the child is invariably non-toxic in appearance [3].

Although there are no specific diagnostic lab tests, blood tests may reveal a mild leukocytosis, lymphocytosis, thrombocytosis, raised C-reactive protein and ESR. Skin biopsy shows characteristic leukocytoclastic vasculitis (Fig. 5) of small dermal blood vessels and on direct immunofluorescence vascular deposits of C1q, C3, fibrinogen and immunoglobulins can be seen [1–3].

The differential diagnoses to consider in AHOI are:

- periorbital cellulitis
- odontogenic abscess
- Henoch-Schönlein purpura

Fig. 4 Generalised purpuric rash over inner thigh

Fig. 5 Skin biopsy showing characteristic leukocytoclastic vasculitis of small dermal blood vessels

- allergies
- meningococemia
- septicaemia
- erythema multiforme
- angioedema
- drug eruptions
- nephrotic syndrome

AHOI is self-limiting with spontaneous recovery occurring within a few days to weeks. Supportive therapy and symptomatic relief is advised and there is no role for steroids & antihistamines [1].

Conclusion

AHOI is likely to be more common than appreciated [3]. It is self-limiting, with no clear cut diagnostic markers, but can be mistaken for other more serious diagnoses and

perhaps over-treated. After thorough investigation, AHOI should be considered in the differential diagnosis of unexplained, dramatic facial/orbital swelling in an otherwise well child.

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