Anales de Pediatría 98 (2023) 483–485

analesdepediatría

www.analesdepediatria.org

IMAGES IN PAEDIATRICS

Remember bradykinin-induced angioedema — an unforgettable image

Recuerde el angioedema inducido por bradicinina: una imagen inolvidable

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Received 22 August 2022; accepted 25 November 2022
Available online 15 May 2023

A boy aged 4 years presented to the emergency department (ED) with exuberant facial swelling of 24 hours duration with progressive worsening. He had been evaluated earlier at a different ED, where he received oral antihistaminic and steroid drugs without subsequent improvement. The physical examination revealed pronounced oedema in the eyelids, lips and nose (Fig. 1). Due to a strong suspicion of bradykinin-induced angio-oedema, C1 inhibitor (C1-INH) was administered through the intravenous (IV) route (twice), followed by IV aminocaproic acid and subcutaneous icatibant (twice) due to a lack of response. A review of the medical history revealed a previous diagnosis of acquired angioedema due to C1-INH deficiency following several episodes of feet swelling. In a few hours, the patient exhibited slow but progressive improvement (Fig. 2). At present, the patient is in treatment with tranexamic acid for prophylaxis and on-demand icatibant.

Angio-oedema and C1-INH deficiency are potentially fatal diseases characterised by oedema of the skin or mucosae involving chiefly the skin, gastrointestinal tract and upper respiratory tract.[1–3] Early treatment reduces the duration of the attack, so it is crucial to identify patients who could benefit from it (Fig. 3).[1–3] In the case of our patient, the delay in the initiation of appropriate treatment could explain his delayed response, which underscores the importance of the identification and treatment of angio-oedema in order to prevent the associated morbidity and mortality.

Ethical considerations

The authors obtained the written informed consent of the legal guardians of the patient.

DOI of original article: https://doi.org/10.1016/j.anpedi.2022.11.007
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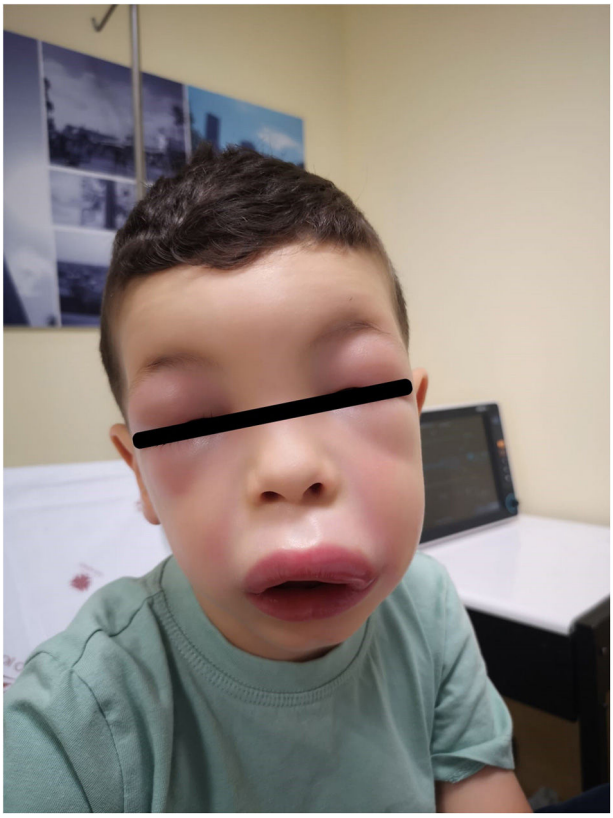

Figure 1 Initial presentation at the emergency department during the acute exacerbation.

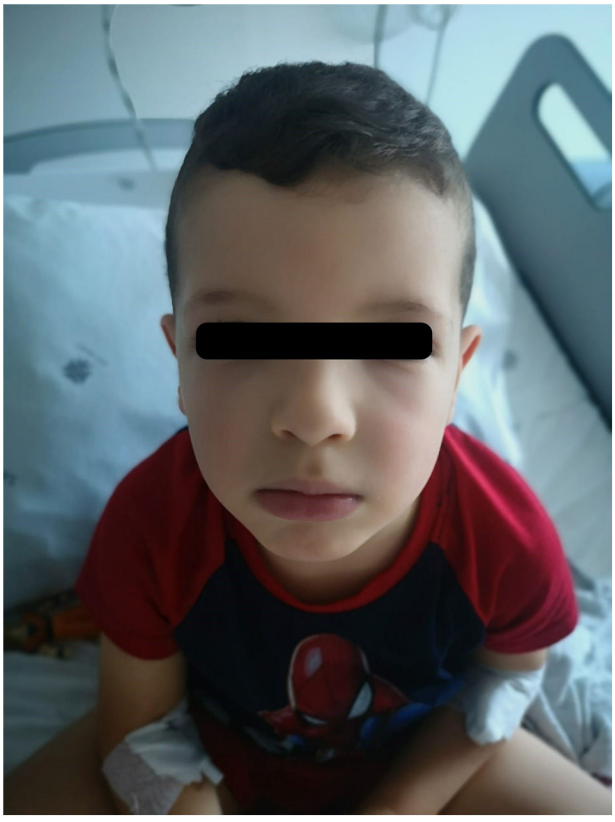

Figure 2 The patient at discharge, after the administration of IV C1 inhibitor, IV aminocaproic acid and subcutaneous icatibant.

<table>
<tr><td>Bradykinin-induced angio-oedema. Suspect in the case of:
1. Recurrent cutaneous/mucosal inflammation in any location (chiefly face, extremities, genitals, intestinal mucosa, oropharynx or larynx), without urticaria and in the absence of any other identifiable cause.
2. HAO in first-degree relative</td><td rowspan="2">Diagnostic tests
• Measurement of serum level of C1-INH, C1-INH activity level and serum level of C4 (if results are indicative of HOE type ½, repeat tests in 1-3 months for confirmation)

Acute treatment for any form of HAO:
First-line treatment:
• Icatibant (if poor response, repeat in 6 hours) or C1-INH (in the case of HAO type 1/2)
Second-line treatment (antifibrinolytics):
• Tranexamic acid (if poor response, repeat in 6 hours) or C1-INH (in the case of HAO type 1/2)
Do not use steroids, antihistamines or epinephrine, as they are not effective</td></tr>
<tr><td>Bradykinin-induced angio-oedema. Different types:
1. HAO type 1: with deficient C1-INH
2. HAO type 2: with dysfunctional C1-INH
3. HAO-nlC1-INH: HAO with normal levels of C1-INH associated with 6 different variants*</td></tr>
<tr><td>Urgent treatment in case of:
1. Inflammation of the larynx, face or neck
2. Abdominal attacks
3. Disabling attacks</td><td>Plasma-derived C1-INH is recommended for first- and second-line prophylaxis in paediatrics. Short-term prophylaxis is recommended before medical, surgical or dental procedures or other events that may trigger attacks. Individualise long-term prophylaxis: first-line, pdC1-INH; second-line, antifibrinolytics.</td></tr>
<tr><td colspan="2">C1-INH, C1 inhibitor; HAO, hereditary angio-oedema; pdC1-INH, plasma-derived C1-INH. Normal ranges: C1-INH level (17.4-24.4 mg/dL); C1-INH activity (70%-130%); C4 level (6-96 mg/dL). Dosage for acute treatment: subcutaneous icatibant 10 mg (weight 12-25 kg), 15 mg (26-40 kg), 20 mg (41-50 kg), 25 mg (51-65 kg) or 30 mg (> 65 kg); IV C1-INH 20 U/kg; IV tranexamic acid 7-10 mg/kg; IV e-aminocaproic acid 100 mg/kg.
*Changes in the factor XII gene (HAEFXII), angiopoietin-1 gene (HAE-ANGPT1), plasminogen gene (HAE-PLG), kininogen 1 gene (HAE-KNG1), myoferlin gene (HAE-MYOF) and heparan sulphate 3-O-sulphotransferase 6 gene (HAE-HS3ST6).
References 1 and 3.</td></tr>
</table>

Figure 3 Management of patients with hereditary angioedema.

References

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