

# Recurrent hemifacial oedema

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## DESCRIPTION

A 3-year-old boy, with no relevant family/personal history, presented at the emergency department due to recurrent left hemifacial erythematous oedema and feverish peak. The mother reported two similar episodes in the previous months, interpreted as periorbital cellulitis, with resolution with systemic antibiotic and oral corticosteroid. She denied fever, weight loss or other constitutional symptoms.

At examination presented skin paleness; soft, non-tender swelling and erythema of the left periorbital region (Figures 1); shotty, non-tender, well defined, smooth generalised lymphadenopathy (cervical, submaxillary, axillary, inguinal); firm and non-tender splenomegaly palpable 2 cm below the costal margin. Ophthalmological examination and funduscopy were normal.

Laboratory tests revealed a normocytic and normochromic anaemia (haemoglobin of 8.4 g/dL), leucocytosis (28100 WBC/ $\mu$ L) with 77% of lymphocytes, 137.000/ $\mu$ L platelets, normal liver function enzymes and negative serology for Epstein-Barr virus. Peripheral blood smear revealed 44% of blasts; flow cytometry was compatible with B cell acute lymphoblastic leukaemia (ALL). Abdominal

ultrasound identified homogeneous hepatosplenomegaly and visceral lymphadenopathies; chest X-ray was normal.

Ophthalmological and periorbital manifestations have been frequently reported during the evolution of ALL (as much as 90%), either by leukaemic infiltration or secondary to haematological derangements (such as anaemia, thrombocytopenia, leucopenia and hyperviscosity).<sup>1 2</sup> Recurrent hemifacial oedema has not been described as a presenting symptom of ALL; however, it has been reported in the superior vena cava syndrome.<sup>3</sup> We hypothesise that the unilateral and fluctuating character of the oedema may have been related to lymph node compression of peripheral lymphatic or vascular structures. Corticosteroid treatment might have contributed to the transient remission in the previous episodes.

**Contributors** MM: bibliographic research, data collection, analysis and interpretation of data and main role in writing the manuscript, and approval of the final version of the article. NS: writing the manuscript and approval of the final version of the article. SG and CC: substantial intellectual contribution, participation in the analysis and interpretation of data, critical review of content and approval of the final version of the article.

**Competing interests** None declared.



Figure 1. Oedema and erythema of the left periorbital region.



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## Learning points

- ▶ The presenting symptoms of paediatric cancer are often non-specific and may be difficult to distinguish from common and self-limiting childhood illnesses.
- ▶ Recurrent hemifacial oedema, although not a common manifestation of acute lymphoblastic leukaemia was the presenting sign of the disease in this case.

**Patient consent** Obtained from guardian.

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