Isolated Acute Hemorrhagic Edema of the Eyelids

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ABSTRACT

Perivascular epithelioid cell tumors (PEComas) are rare tumors which can arise in almost every body site. Little is still known about these tumors because of the limited number of cases described by far and the short follow-ups. Diagnosis, based on histopathological and immunohistochemical features, is not so straightforward owing to their rarity. Prediction of clinical behaviour which can be benign, locally aggressive or purely malignant, is not without flaws. A refinement in the prognostic criteria is desirable. Even if surgery is considered the mainstay of therapy of both primary PEComa and local recurrences/metastases, doubts still exist regarding the other treatment modalities such as chemotherapy and immunotherapy. The rarity of this neoplasm prompted us to report a case we observed. A left lung PEComa was diagnosed in a 70-year-old lady that we treated with upper left lobectomy. Our aim is to add our experience to the present body of literature and contribute in the understanding of PEComa neoplasms.

Key words: Hemorrhagic edema; Vasculitis; Infancy; Pediatrics

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Abbreviations

WBC: white blood cells; AHEI: acute hemorrhagic edema of infancy

CASE REPORT

We evaluated a 34-day-old girl for an isolated acute hemorrhagic edema of the eyelids (Figure 1) and hyperpyrexia. She had been evaluated two days before for an upper respiratory tract infection. Her past history was also relevant for a successfully treated early neonatal streptococcal B sepsis.

Lab tests showed leukocytosis (WBC 145,600/mm³, reference range: 5,000-19,500/mm³), and slightly raised C-reactive protein (1.9 mg/L, reference range: 0-0.5 mg/L).

The presence of bilateral eyelid swelling and purpura in a well-appearing child raised our clinical suspicion for an isolated variant of acute hemorrhagic edema of infancy (AHEI). An ocular swab and hemoculture turned out negative.

24 hours after admission, fever dropped abruptly and eyelid edema reduced. 72-hours control confirmed regression of the hemorrhagic edema in a well-appearing and thriving toddler (Figure 2).
DISCUSSION

AHEI is characterized by the triad of fever, edema, and annular “target-shaped” purpura over the face, ears, and extremities of children younger than 3 years of age[1-5].

Onset is dramatically fast: after 24-48 hours, bruises and purple ecchymoses are fully visible in the interested areas. Once thought to be a variant of Henoch-Schönlein purpura, AHEI is now considered a distinct vasculitis, because of the infrequency of both visceral involvement and Immunoglobulin A skin depositions[5], as well as a better prognosis.

The specific etiology of AHEI is unknown, but may be related to viral infections, medications, and immunizations[4,5].

To date, few cases of isolated eyelids AHEI are described, none in children younger than 2 months of age.

REFERENCES