syndrome, and the patient was referred for neurologic consultation.

PHACE syndrome is an acronym that was coined in 1996 to describe this constellation of clinical features,[1] with consensus-derived diagnostic criteria that were updated in 2016.[2] The syndrome is variably referred to as PHACES, to account for the midline chest and abdominal anomalies (including sternal pits and clefts and supraumbilical raphe). Ventral midline skin blanching also has been described.[3] Although the infantile hemangiomas in PHACE syndrome are typically large and segmental (lesions that cover a larger anatomic territory of the face or body), they can be more subtle, as demonstrated in our patient. However, their multifocal nature, in conjunction with the midline developmental defects, heightened our concern for this syndrome, prompting further evaluations that confirmed the diagnosis. Given that the cerebral arteriopathy in PHACE syndrome may predispose patients to arterial ischemic stroke and impact risk stratification of infantile hemangioma treatment with beta blockers, as well as other potential long-term morbidities, prompt recognition and diagnostic confirmation are vital. We present this patient to highlight the importance of meticulous skin examination in this setting, including evaluation for subtle midline developmental anomalies, which may serve as a critical diagnostic finding. ■

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Isolated Forehead Swelling

A previously healthy 3-year-old girl presented to the dermatology department with a 3-week history of swelling of the right forehead. The lesion was a nonpainful, nonfluctuant, nonpulsating, indurated mass of about 3 × 3 cm, increased in size over the past 2 weeks, and with overlying skin eventually turned purple (**Figure 1**, A). Ultrasound examination revealed a solid hyperechoic subcutaneous lesion with enhanced Doppler signal, intact underlying osseous plate, and ipsilateral submandibular and cervical lymphadenopathy. Abdominal ultrasound and chest radiogr aph were normal. Skull radiograph ruled out osteolytic bone damage. Laboratory tests showed isolated high white blood cell (WBC) count (16 600/mm^3: neutrophils 10 800, lymphocytes 3700, monocytes 880, eosinophils 1000) with no elevation of inflammatory markers, and lactate dehydrogenase, ferritin, and neuronal specific enolase within the normal range for age. No peripheral blood smear was obtained on that occasion. In the hypothesis of a skin infection, a course of oral amoxicillin-clavulanate was started.

Ten days later, the patient was brought to the emergency department for high fever and severe back pain: the skin lesion had further increased in size despite the antibiotic therapy. Repeated blood tests showed WBC count increased up to 25 000 cells/mm^3 (neutrophils 13 000, lymphocytes 6790, monocytes 3910, eosinophils 1000) with mildly elevated C-reactive protein (41 mg/L). A peripheral blood smear showed the presence of 47% monocytic-like blasts (**Figure 2**, A). Nevertheless, the bone marrow aspirate displayed a predominance of lymphoid blasts (**Figure 2**, B), and the immunophenotypic markers confirmed the diagnosis of common B-cell acute lymphocytic leukemia (ALL). A leukemia cutis with newly onset systemic ALL was diagnosed. Steroid prephase was started, and the mass completely resolved by day 8 (**Figure 1**, B).

Leukemia cutis consists of the infiltration of lymphoid or myeloid blasts into the epidermis, dermis, or subcutaneous tissues. It can manifest as petechiae, purpura, macules, papules, patches, plaques, or nodules, mainly involving the head and the lower extremities. Leukemia cutis is most commonly associated with acute myeloid leukemia, and in this case, it is specifically labeled as myeloid sarcoma,

J Pediatr 2021;232:300-2.

https://doi.org/10.1016/j.jpeds.2021.01.001

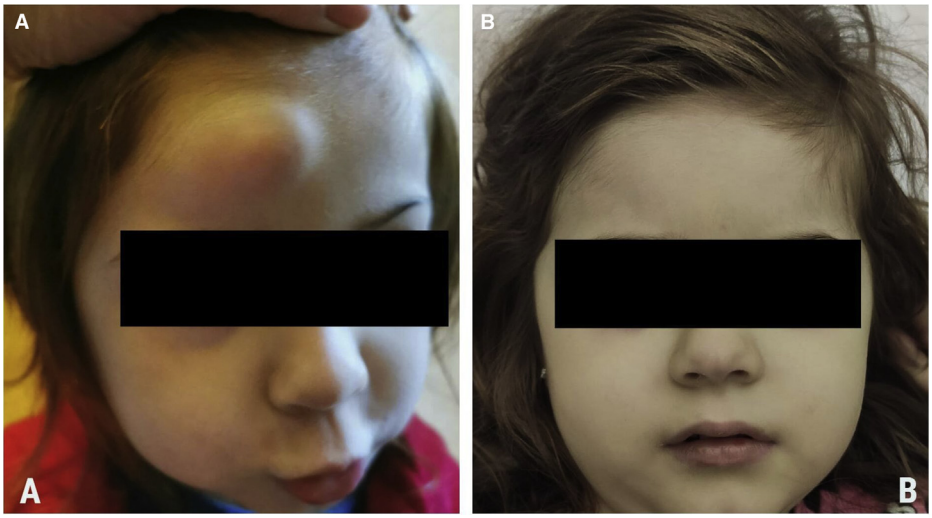

Figure 1. **A,** Right forehead swelling. **B,** Complete resolution of the mass after proper steroid prephase.

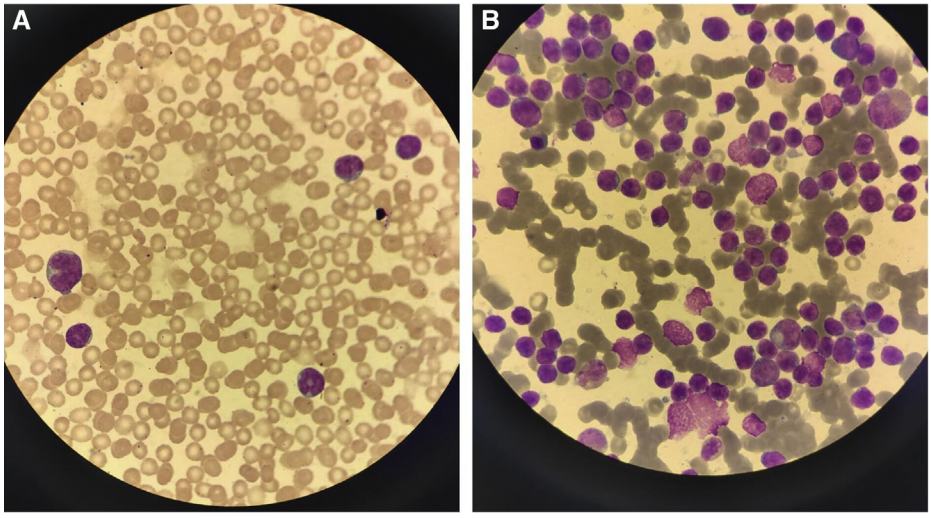

Figure 2. **A,** Peripheral blood smear showing monocytic-like blasts. **B,** Bone marrow aspirate showing lymphoid blasts.

granulocytic sarcoma, or extramedullary acute myeloid leukemia tumor.[1] However, it has also been described in association with ALL.[2] Although generally concomitant or preceded by the occurrence of signs and symptoms of systemic involvement, leukemia cutis can also rarely represent the very first presenting sign.[3]

The differential diagnosis of subcutaneous swelling in children include infective (eg, Pott puffy tumor), vascular (eg, lymphangiomas), hematologic (eg, hemophilia), neoplastic (eg, Langerhans cell histiocytosis, neuroblastoma), and cutaneous disorders (eg, epidermoid/dermoid cyst and giant pilomatrixoma). Peripheral blood smear, bone marrow aspirate, and skin biopsy should be performed in the suspect of leukemia cutis. In this case, the association of an undefined, subcutaneous lesion, unchanged despite antibiotics, yet turning into a severe systemic disease with high WBC and disproportioned inflammatory markers, suggested a hematologic malignancy. ■

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