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IMAGES

Cutaneous ALK-positive histiocytosis

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A 2-year-old female presented with a single 1.2-cm nodular scalp lesion that was surgically resected. Histopathologic examination revealed a dermal infiltrate of large mononuclear cells, with abundant eosinophilic cytoplasm, and lobulated/irregularly shaped nuclei with fine chromatin (Fig. 1a–d). Immunohistochemical analysis demonstrated negative staining for CD1a, strong staining for lysozyme, and prominent cytoplasmic staining for ALK (Fig. 1e–g); there was also strong staining for CD68 (PG-M1) and negative staining for CD30 and S100 (not shown). Fluorescence in situ hybridization (FISH) confirmed an *ALK* rearrangement in 68.5% of 100 interphase tumor cell nuclei (Fig. 1h). The patient had no clinical evidence of recurrent disease 2 years status post-surgical resection.

ALK-positive histiocytosis occurs most often in the pediatric age group, especially at less than 3 years of age [1–4]. A few cases have occurred in adults [2, 5]. Patients may present with a localized lesion (skin, brain, breast, or appendix) or with disseminated disease (liver, spleen, bone marrow, and skin) [1–5]. Most patients have had a favorable outcome after surgical resection of a localized mass or following chemotherapy or ALK inhibitor treatment for localized or disseminated disease [1–5].

Based on variability in tumor cell morphology and immunophenotype, the differential diagnosis for ALK-positive histiocytosis may include juvenile xanthogranuloma, Rosai-Dorfman disease, Langerhans cell histiocytosis, and Erdheim-Chester disease [2–4]. For tumors that have been studied, *KIF5B* is the most common gene partner for *ALK* [2, 4, 5].

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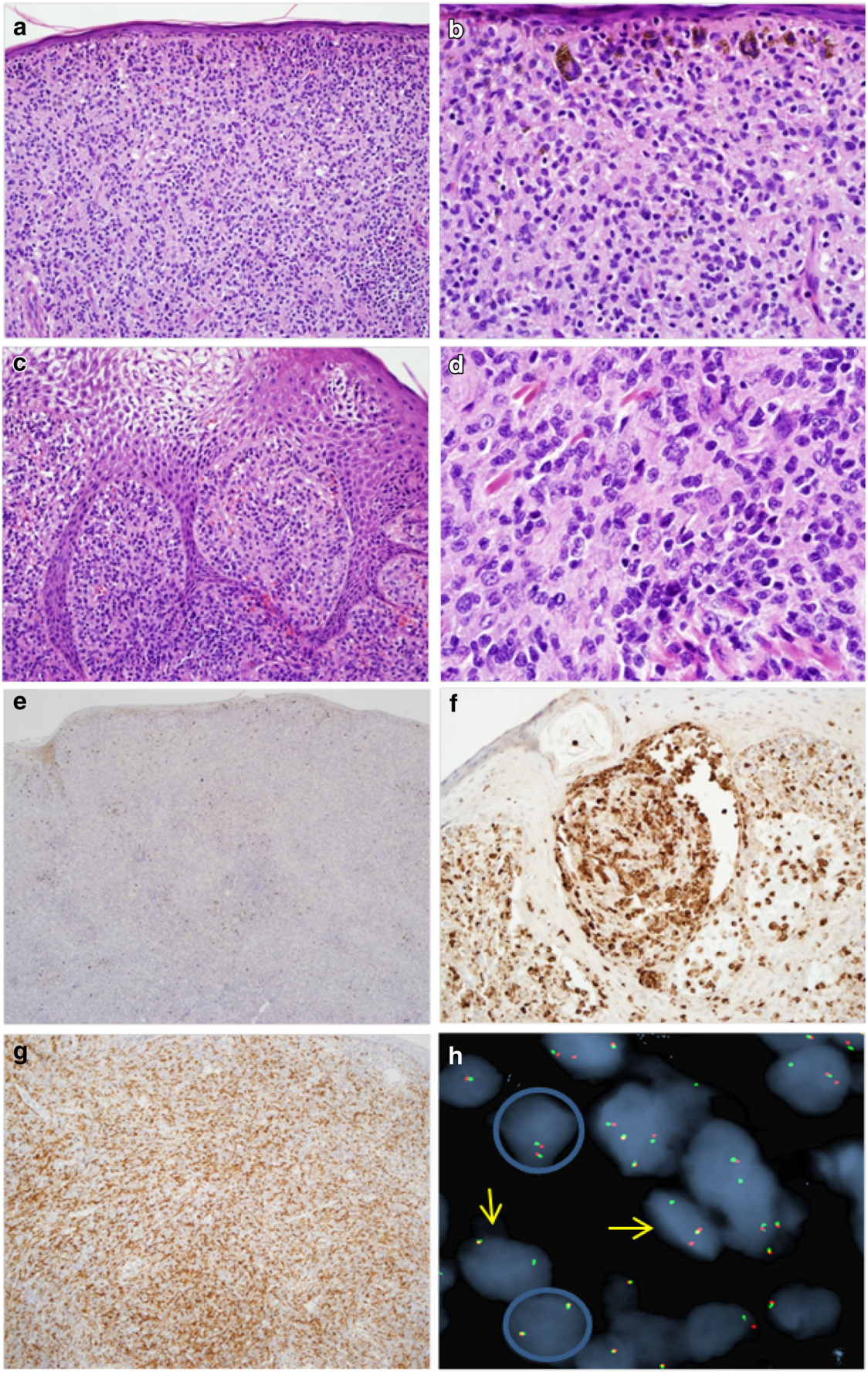
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◀ **Fig. 1** Histologic sections demonstrate a dermal infiltrate composed of large mononuclear cells with irregularly shaped nuclei and abundant eosinophilic cytoplasm (**a**–**d**). The lesional cells are negative for CD1a (**e**); however, strong cytoplasmic reactivity for lysozyme (**f**) and cytoplasmic expression for ALK (**g**) are noted. Gene rearrangement is confirmed by FISH with an *ALK* break-apart probe. The arrows indicate cells with an *ALK* rearrangement (one red, green, and yellow signal), while the circled cells represent no gene rearrangement (2 yellow signals; **h**)

Declarations

Conflict of interest The authors declare no competing interests.

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