PAEDIATRIC DERMATOLOGY

A CASE OF AN ALK-POSITIVE, ATYPICAL SPITZOID TUMOR IN A 2-YEAR-OLD GIRL

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Background: Spitz nevi are benign in nature and rarely progress to melanomas. Atypical Spitz tumors pose a diagnostic challenge as they behave in an unpredictable manner. Research into the genetic milieu of spitzoid tumors has revealed many genetic aberrations that may have diagnostic and prognostic implications.

Observation: We report a case of a 2-year-old Hispanic female with no personal or family history of melanoma who presented with an enlarging nodule on the chin. Upon initial presentation, the mother reported trauma to the chin 3 months prior; physical examination revealed a 7 mm, dome-shaped, red papule on the chin with regularly distributed globules on dermoscopy. Two months later, the lesion enlarged in size; physical examination revealed a 1.4 cm x 1.2 cm exophytic, deep red nodule on the chin remarkable for irregularly distributed brown globules on dermoscopy. A shave biopsy of the nodule was performed and histopathologic evaluation revealed multiple large nests of epithelioid and spindle-shaped melanocytes, confluent at all levels with numerous mitotic figures. This was interpreted as an atypical compound spitzoid melanocytic neoplasm with inflammation. Immunohistochemistry was notable for anaplastic lymphoma kinase (ALK) overexpression. Additional molecular studies with amplicon-capture next generation sequencing was negative for 98 cancer-related mutations. The results were suggestive of an ALK-positive, atypical spitzoid melanocytic proliferation.

Key message: Genomic aberrations in atypical Spitz tumors are diverse and infrequently observed in other melanocytic neoplasms. ALK-positive Spitz tumors are more often reported in younger individuals and histopathologically feature fascicles of larger epithelioid melanocytes, which may be explained by activation of the PI3K/AKT/mTOR pathways. These tumors tend to behave in an indolent manner but some rarely metastasize, requiring systemic treatment. Given recent advances in targeted therapy, ALK inhibitors may pose a therapeutic option for these neoplasms.