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Diagnosis, Treatment, and Prognosis in Cutaneous Anaplastic Lymphoma Kinase-Positive Anaplastic Large Cell Lymphoma: A Narrative Review Apropos of a Case

Authors : Laura Gleason, Sahithi Talasila, Lauren Banner, Ladan Afifi, Neda Nikbakht

Abstract : Primary cutaneous anaplastic large cell lymphoma (pcALCL) accounts for 9% of all cutaneous T-cell lymphomas. pcALCL is classically characterized as a solitary papulonodule that often enlarges, ulcerates, and can be locally destructive, but overall exhibits an indolent course with overall 5-year survival estimated to be 90%. Distinguishing pcALCL from systemic ALCL (sALCL) is essential as sALCL confers a poorer prognosis with average 5-year survival being 40-50%. Although extremely rare, there have been several cases of ALK-positive ALCL diagnosed on skin biopsy without evidence of systemic involvement, which poses several challenges in the classification, prognostication, treatment, and follow-up of these patients. Objectives: We present a case of cutaneous ALK-positive ALCL without evidence of systemic involvement, and a narrative review of the literature to further characterize that ALK-positive ALCL limited to the skin is a distinct variant with a unique presentation, history, and prognosis. A 30-year-old woman presented for evaluation of an erythematous-violaceous papule present on her right chest for two months. With the development of multifocal disease and persistent lymphadenopathy, a bone marrow biopsy and lymph node excisional biopsy were performed to assess for systemic disease. Both biopsies were unrevealing. The patient was counseled on pursuing systemic therapy consisting of Brentuximab, Cyclophosphamide, Doxorubicin, and Prednisone given the concern for sALCL. Apropos of the patient we searched for clinically evident, cutaneous ALK-positive ALCL cases, with and without systemic involvement, in the English literature. Risk factors, such as tumor location, number, size, ALK localization, ALK translocations, and recurrence, were evaluated in cases of cutaneous ALK-positive ALCL. The majority of patients with cutaneous ALK-positive ALCL did not progress to systemic disease. The majority of cases that progressed to systemic disease in adults had recurring skin lesions and cytoplasmic localization of ALK. ALK translocations did not influence disease progression. Mean time to disease progression was 16.7 months, and significant mortality (50%) was observed in those cases that progressed to systemic disease. Pediatric cases did not exhibit a trend similar to adult cases. In both the adult and pediatric cases, a subset of cutaneous-limited ALK-positive ALCL were treated with chemotherapy. All cases treated with chemotherapy did not progress to systemic disease. Apropos of an ALK-positive ALCL patient with clinical cutaneous limited disease in the histologic presence of systemic markers, we discussed the literature data, highlighting the crucial issues related to developing a clinical strategy to approach this rare subtype of ALCL. Physicians need to be aware of the overall spectrum of ALCL, including cutaneous limited disease, systemic disease, disease with NPM-ALK translocation, disease with ALK and EMA positivity, and disease with skin recurrence.

Keywords : anaplastic large cell lymphoma, systemic, cutaneous, anaplastic lymphoma kinase, ALK, ALCL, sALCL, pcALCL, cALCL
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