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Unusual Presentation of T-cell non-Hodgkin Lymphoma with Multiple Skin Nodules

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Abstract

Non-Hodgkin lymphoma is very uncommon in infancy and skin as the primary site of involvement in Non-Hodgkin lymphoma is rarely encountered. We describe a 10-month-old infant with T-cell Non-Hodgkin lymphoma who presented with multiple skin nodules as the predominant feature of her disease. The clinical manifestations, treatment strategy and disease outcome are reviewed.

Key words: Non-Hodgkin lymphoma, T-cell, skin nodules, infancy.

Introduction

Non-Hodgkin lymphoma (NHL) is an uncommon malignancy in children younger than 5 years old. Approximately, half of the cases of childhood NHL are derived from cells of T-cell lineage [1]. The most common clinical features of T-cell NHL are anterior mediastinal mass and upper torso lymphadenopathy. Skin as the primary site of involvement is rare in T-cell NHL, except in anaplastic large cell variant [2]. We report a 10-month-old infant, who is a case of T-cell NHL with unusual presentation of multiple skin nodules.

Case report

A 10-month-old female infant was first brought to the emergency ward due to severe abdominal protrusion and respiratory distress. Her past medical history was unremarkable. In physical examination, she was afebrile but tachypneic and tachycardic. Hepatosplenomegaly or peripheral lymphadenopathy was not detected.

Abdominal sonography showed bilateral enlarged kidneys with increased cortical echogenicity and distorted echotexture and possibility of Wilms' tumor was proposed. Due to her poor general condition, she was transferred to pediatric intensive care unit and received one dose chemotherapy including vincristine, cyclophosphamide and adriamycin. Two days later, she was transferred to pediatric oncology ward for further evaluation. Abdominal CT scan was performed which showed mildly enlarged kidneys, but no mass or intra-abdominal lymph node was detected. Other workups including chest x-ray, complete blood count, 24 hr urine for measurement of vanilmandelic acid was performed. Bone marrow aspiration and biopsy were normal, except a high level of lactate dehydrogenase. She was discharged with close follow-up, but did not come back until 3 months later with multiple large, firm, non-tender masses on the scalp, forehead, chest wall, trunk and lower extremities (Figure 1.A).

Histologic examination of the tissue biopsy specimen revealed infiltration of monomorphic small cell malignant tumor with soft tissue, muscle and subcutaneous tissue involvement (Figure 2.A). Immunohistochemical staining was in favor of T-cell NHL (positive for leukocyte common antigen (LCA) and CD3) (Figure 2.B). Sections stained for CD20, cytokeratin, terminal deoxynucleotidyl transferase (TdT) and anaplastic lymphoma kinase antibody (ALK1) were negative. Polymerase chain reaction for detection of Epstein-Barr virus was negative in neoplastic cells.

Chemotherapy was started according to non-B-cell lymphoma protocol (BFM-NHL protocol, P. Lanzkovski, 2005, 4th edition). After two courses of treatment, all of skin nodules disappeared completely (Figure 1.B). Her treatment is going on

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and she is in good general condition and regular follow up is performed at the time of this report.

Discussion

NHL is uncommon in children younger than 5 years in whom, it accounts for 3% of cancers. Pediatric NHL differs from adult disease in that, they are almost all high grade, have a diffuse growth pattern and commonly involve extranodal sites [3]. The most common extranodal sites for NHL are the gastrointestinal tract and nasopharynx followed by skin, brain, bone, thyroid, breast, lung and testis [4].

Skin as the primary site of involvement in NHL is rare and accounts for only 1-3% of cases, primarily in advanced disease [5].

Skin is more frequently involved in CD30-positive anaplastic large cell lymphoma and precursor B-cell lymphoblastic lymphoma rather than in other childhood NHLs. Malignant skin lesions, presenting as skin nodules, are very rare in children and highly uncharacteristic. It can be seen in various hematologic disorders including neuroblastoma, rhabdomyosarcoma, leukemia and lymphoma. The final diagnosis is often delayed and requires biopsy for pathology confirmation.

The predominant location of the cutaneous nodules has been previously reported to be on the scalp and forehead, followed by neck, trunk and extremities [1,3,5-8]. This was the same as what was observed in our patient with multiple skin nodules on the scalp, forehead, trunk and feet.

Although in our patient, we could not verify any other location of lymphomatous infiltration except skin nodules; we think that it was a case of secondary cutaneous lymphoma. In fact, it seems that the enlarged kidneys detected by ultrasonography at presentation, were the systemic manifestations of NHL and skin nodules which appeared about 3 months later, were secondary dissemination of the tumor to the skin.

As the published data on cutaneous NHL in children are very scant, it is difficult to elaborate a strategy of treatment.

These children are often treated like all other extranodal NHL cases; with multi-drug chemotherapy [9]. We treated our patient with systemic chemotherapy according to BFM-NHL protocol for non-B-cell NHL.

In contrast to what Reich described about the poor outcome of children with NHL and secondary cutaneous involvement,9 our patient had dramatic response to chemotherapy. She is on maintenance therapy and is in complete remission after 12 months regular follow up. The dramatic response of our case to chemotherapy resulting in total disappearance of all of the skin nodules (Figure 3), supports the proposal that this condition might be highly curable despite diffuse dissemination at primary presentation.

Conclusion: T-cell NHL with multiple skin nodules should be considered as a rare presentation in infancy and systemic chemotherapy should be started as soon as possible.

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Figure 1.A



Figure 2.A

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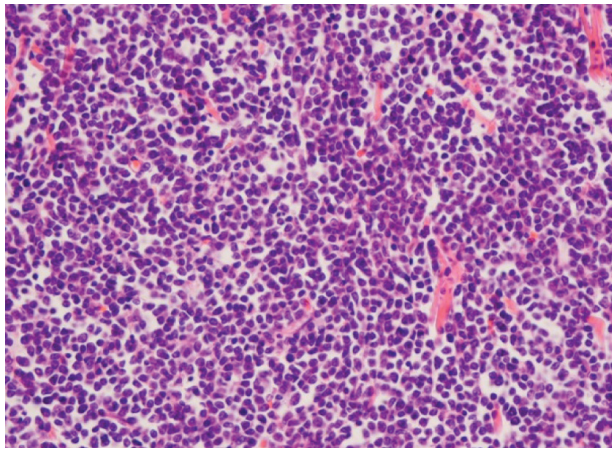


Figure 1.A

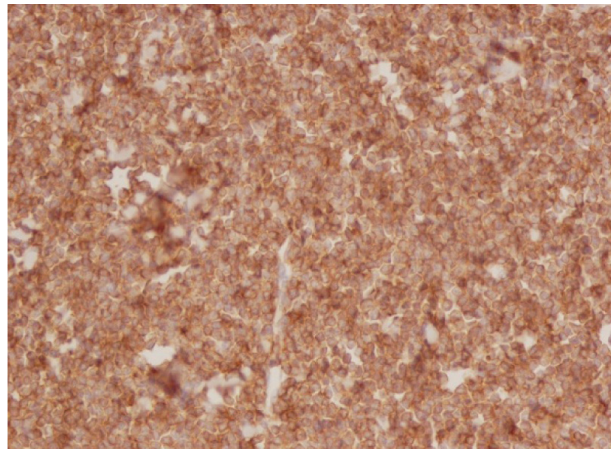


Figure 2.A

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