SKIN CANCER (OTHER THAN MELANOMA)

A RARE CASE OF MULTISYSTEMIC LANGERHANS CELL HISTIOCYTOSIS MIMICKING DIFFUSE HEMANGIOMATOSIS

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Background: Langerhans cell histiocytosis (LCH) is a rare disorder characterized by abnormal proliferation of histiocytes, commonly presents as acute, disseminated multisystemic disease. Skin manifestation of LCH is variable and the most common presentation is multiple, rose-yellow, scaly papules located on the scalp, face and trunk, mimicking seborrheic dermatitis. In the last two decades, diffuse neonatal hemangiomatosis (DNH) like eruptions are described in congenital self healing reticulohistiocytosis (CSHR)- a benign subtype of LCH-, however few cases of multisystemic LCH demonstrated DNH as skin manifestation. Here we present a rare case of LCH mimicking DNH with multisystemic involvement.

Observation: A 4 -month old female was referred to outpatient clinic with erythematous rash mostly on the trunk, associated with on-and-off fever for 2 months. On physical examination, multiple, discrete, red-purple papules ranging in size between 0.3-0.8 cm, located mainly on the trunk, were identified. She had also rose-yellow scaly papules on the head and face. Dermoscopy revealed red-blue lacunas mimicking hemangiomas on the trunk and scaly yellow papules resembling seborrheic dermatitis on head and face. Histopathology of the lesions showed infiltration of the papillary dermis with histiocytes with notched nucleus and abundant eosinophilic cytoplasm. Cells were CD1a, S100 and langerin positive. Patient was referred to pediatric oncology department, sequential workup showed involvement of the lungs and thymus.

Key message: LCH is a proliferative disorder that can mimic childhood inflammatory and neoplastic conditions. Differential diagnosis include infantile hemangiomas, hemangioendoteliomas, lymphatic malformations, infantile myofibromatosis, histiocytomas as benign conditions. Fibrosarcoma, PNET and other sarcomas are most common malignant mimickers. In conclusion, our case illustrates the importance of early excision of fully formed vascular lesions with sudden appaerance in infants, that can harald malignancy.