Congenital Langerhans Cell Histiocytosis Presenting as Neonatal Papulovesicular Eruption - Rapid Recurrence with Multisystem Involvement after Spontaneous Regression of Primary Lesion

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Abstract

Congenital self-healing Langerhans cell histiocytosis (CSHLCH) is a rare type of Langerhans Cell Histiocytosis, presenting at birth or during the neonatal period. It is usually characterized by the eruption of multiple, discrete, red-brown papules or nodules which may increase in size and number during the first few weeks of life with spontaneous regression. Congenital LCH has rarely been reported to present as a papulovesicular eruption at birth. Here we describe a male baby presenting with papulovesicular eruption at birth who rapidly developed pulmonary infiltrates and multiple osteolytic lesions in skull and long bones after spontaneous regression of cutaneous lesion.

Key words: Congenital; Histiocytosis; Skin; pulmonary; Bone

Introduction

Langerhans cell histiocytosis (LCH), formerly known as histiocytosis X, is a rare proliferative disorder of bone-marrow-derived antigen presenting cells. LCH in the neonatal period is rare and like LCH in older children and adult, may have a variable presentation and clinical course. Cutaneous changes are the most common initial manifestation in neonates with both Single system-LCH and Multisystem-LCH. Disease course is unpredictable upon diagnosis and ranges from spontaneous healing to chronic course or fulminant deterioration. Reports of congenital cutaneous only LCH describe high tendency to spontaneous regression (so called Hashimoto-Pritzker congenital self-healing histiocytosis). However there is growing evidence that this entity is morphologically indistinguishable from the disseminated form of LCH, and that progression to multi-system disease with fatal outcome is possible. Despite the spontaneous regression of skin lesion in cutaneous self-healing LCH (CSHLCH), close follow up is required for evaluation of systemic dissemination like bone and lung involvement.

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Case Report

A male baby with birth weight of 3.2 kg was born on September 11, 2012 at term by C/S as the third issue of...
At 4 months of age on January 27, 2013 the baby got admitted into the Paediatric Haematology and Oncology unit of Bangabandhu Sheikh Mujib Medical University (BSMMU) with the complaints of respiratory distress for one month, excessive cry on handling and multiple nodular swelling in different areas since 2nd month of his age. Before admission into the paediatric haematology and oncology unit he was treated as a case of pneumonia when he had multiple granular opacities on x-ray chest and histopathology report of left inguinal lymph node biopsy suggestive of viral lymphadenitis.

We examined the baby who was alert, febrile and anicteric but with hurried respiration having no cyanosis or oedema. Lymph nodes were palpable, one in submental, two on both sides of axillary and two on each inguinal area. Size was 1 to 1.5 cm in diameter, discrete nontender not fixed to underlying or overlying structures. Skin survey was normal except for the presence of a scar mark in left inguinal region and few hyperpigmented macules on trunk and buttock. On chest examination the baby had R/R rate 66/min; presence of chest indrawing and sternal recession, breath sound was vesicular with no added sound. Abdomen was slightly distended but soft and nontender with no signs of ascites having just palpable spleen and liver. On locomotor examination there was no bony swelling, deformity or restriction of movement or abnormalities of posture. Joints of different areas were normal, but the bones of his limbs were tender on palpation.

The baby had been re-investigated for infection screening including STORCH, radiological survey of bone and chest and liver function and renal function. Complete blood count, serum electrolyte, creatinine, bilirubin and liver enzymes were normal. The serological test for toxoplasma, rubella, cytomegalovirus, herpes and syphilis were negative. Chest X-ray showed diffuse reticular opacities of the lung parenchyma on both lung fields. Radiological survey of skeletal showed multiple osteolytic lesions on skull and long bones of both lower limbs. Ultrasonography of the abdomen was normal except splenic enlargement.

On such findings a LCH was suspected. Slide from biopsied lymph node was reviewed for second opinion. Review of histopathology revealed that nodal architecture was partially replaced by proliferation of lymphocytes, eosinophils and a good number of histiocytes. Some of histiocytes were atypical looking. A few giant cells were also seen. These cells were S-100 positive on immunohistochemistry. As CD1a not available in our set up so it was not done.
Despite the history of regression of cutaneous lesion, as the disease progresses with involvement of lung and multiple bony sites after a latent period of 1 month, we decided to start systemic chemotherapy and observe him closely. Now the baby is getting weekly injectable vinblastine and daily oral prednisolone.

**Discussion**

Langerhans cell histiocytosis affects all age groups and is characterized by an extremely heterogeneous clinical spectrum, natural course, and outcome. LCH limited to the skin (Cutaneous LCH known as Hashimoto-Pritzker syndrome) and disseminated LCH are the two major forms of the disease found in foetus and neonate. Skin lesions were the main initial manifestation from birth in both cutaneous only LCH and multisystem or disseminated LCH in neonate. The time interval before the appearance of systemic symptoms, if occurs, varies from days to months. Diagnosis of our case as Congenital Cutaneous Self-healing Langerhans cell histiocytosis (CSHLCH) was made on the basis of clinical, radiological features, histopathology, and immunohistochemistry.

Congenital cutaneous LCH was reported for the first time in 1973. But congenital cutaneous Self-healing Langerhans cell histiocytosis (CSHLCH) presenting as papulovesicular eruption has rarely been reported. The characteristic and most interesting feature of the lesion of CSHLCH is their spontaneous involution without treatment usually by 3 months of age leaving behind hypopigmented or hyperpigmented macules. But in our case the disease progressed to involve multiple systems like lung and bone after spontaneous healing of cutaneous lesion.

**Conclusion**

Progression of CSHLCH presenting as papulovesicular eruption to involve multiple systems after spontaneous regression is rare. Neonates diagnosed as congenital cutaneous Langerhans cell histiocytosis should be carefully monitored for progression of the disease.

**References**