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Case Report

Congenital Langerhans cell histiocytosis with hematological manifestations: a rare case

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ABSTRACT

Langerhans cell histiocytosis is a rare disorder of immune cells typically found in skin whose abnormal proliferation leads to inflammation and multi organ damage. It is even rarer in the neonatal population, particularly in preterm neonates. The clinical manifestations of disease could be limited or skin or involve multiple organs to become the more lethal multi systemic LCH. The risk organs include liver, spleen and bone marrow. Immuno histo chemistry using Langerin (CD207) and CD1a provide a definitive diagnosis. The case report is of a preterm neonate with Multisystemic involvement of skin, eyes and bone marrow and aims to highlight the importance of having a high degree of clinical 37 suspicion for early diagnosis and timely treatment of the disease. Despite modern advancements the prognosis for these patients remains bleak.

Keywords: Histiocytosis, Immunohistochemistry, Langerin, Anemia, Thrombocytopenia

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disorder that involves the abnormal proliferation of Langerhans cells, which are specialized immune cells typically found in the skin and involved in antigen presentation. In LCH, these cells accumulate in 52 various tissues and organs, leading to inflammation and subsequent multi organ damage.

LCH is rare in newborns, but it can occur. The overall incidence of LCH in the general population is estimated to be about 1 in 200,000 children annually.[1] Cases of neonatal LCH typically account for a very small fraction of all the pediatric cases, with estimates suggesting fewer than 10% of all LCH cases being diagnosed in infants younger than 1 year, and only a small subset of these is in the neonatal period. The aim is to study the clinical patterns, course, and outcome of neonatal LCH. Here authors present the case of a 32 Week preterm male neonate with findings of LCH at a government hospital in North India.

CASE REPORT

History

The case is of a male neonate born at 32 weeks who presented with a widespread reddish-brown maculopapular rash. He was born to mother, G3P2L1, in a non-consanguineous marriage. There is a history of an elder sibling who passed away from an unknown cause. The mother had gestational diabetes, which was well controlled on Metformin. She presented with preterm premature rupture of membranes and breech presentation, leading to the baby being delivered via cesarean section (LSCS) under dexamethasone coverage.

Physical examination

The baby was born AGA with stable vitals at birth but showed weight loss as the disease progressed. Multiple diffuse maculopapular, hemorrhagic lesions were seen at birth. The lesions were seen on scalp, neck, axillary

region, post auricular area, pinna, front and back of chest and abdomen. Involvement of palms and soles were also noted but mucus membrane was spared. The lesions varied in size with smallest measured at 0.5 mm and largest at 20 mm. The lesions were observed to be firm in consistency and non-blanching. Few were seen as papulonodular lesions with crusting. No hepatosplenomegaly or lymphadenopathy was found. Eye examination of the baby revealed bilateral disc pallor with poorly vascularized central retina and avascular peripheral zone. The BERA and middle ear examination was within normal limits. All other systemic examination was done and found to be consistent with that of a normal preterm neonate. Diffuse maculopapular erythematous rash present over entire body including axilla H & E stain of skin biopsy from sole shows S 100 positive spindle tumour cells with dense chromatin. The baby on day 1 of life presented with widespread maculopapular rash on his body with stable vitals and was shifted to mother's side with spoon feeds.

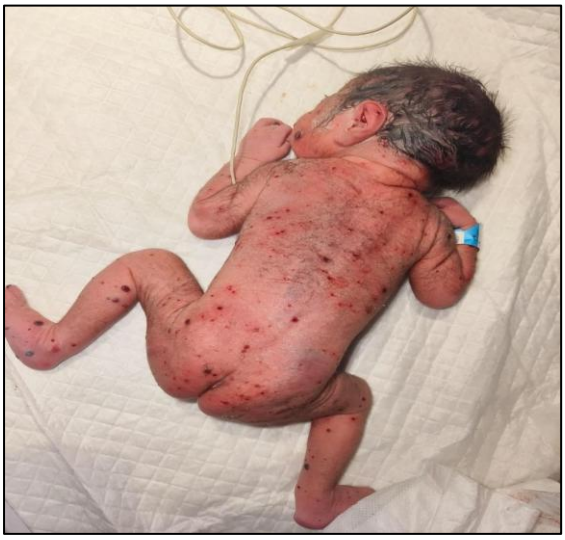

Figure 1: Diffuse maculopapular erythematous rash present over entire body.

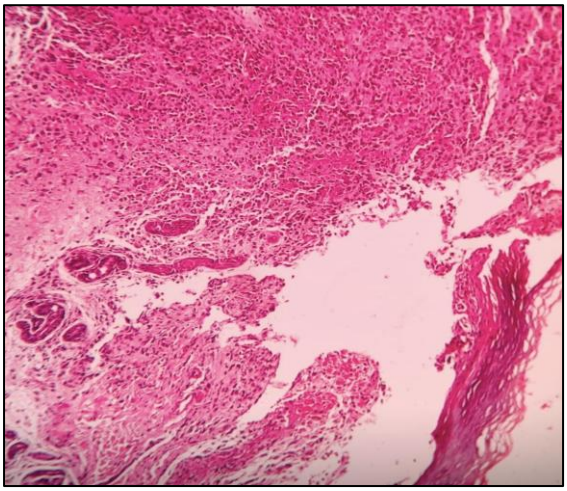

Figure 2: Hematoxylin and eosin 10X.

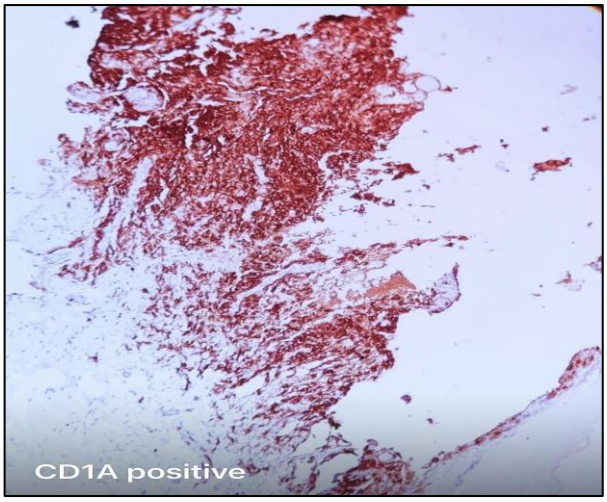

Figure 3: CD1A staining revealed CD1a positive tumor cells.

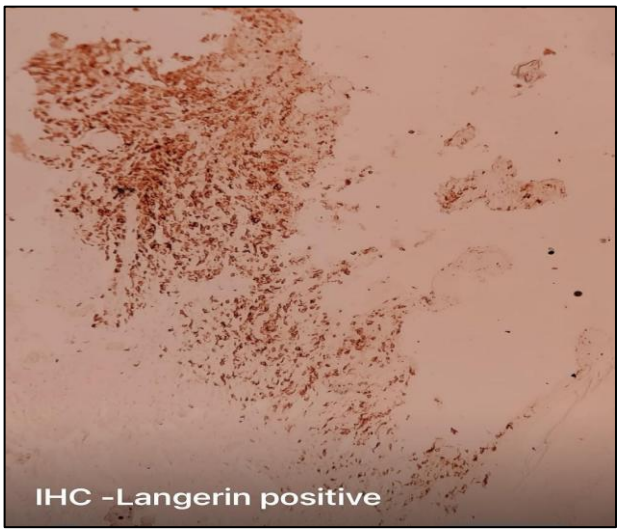

Figure 4: IHC Langerin (CD 207) positive staining of skin biopsy.

The baby's lab results measured over a period of 14 days from birth revealed progressive pancytopenia including anemia and severe thrombocytopenia which led to recurrent episodes of bleeding. PT INR was highly raised. The lipid profile, LFT, Thyroid Function test and urinalysis were normal but with slightly raised urea, creatinine levels. VDRL and TORCH panel done at birth were negative. The skeletal survey, CXR and USG whole abdomen were also within normal limits.

Culture of blister fluid was sterile and tissue diagnosis taken on Day 5 of life revealed S 100 positive tumour cells with spindle and curved nuclei containing dense chromatin. Further IHC CD1a and Langerin (CD207) were performed and a diagnosis of LCH was made. The baby started deteriorating on Day 9 of life with decreased oral intake and sluggishness. The baby was shifted back to NICU and started on IV fluids and antibiotics anticipating sepsis in view of raised CRP levels. A septic

screening performed turned negative. The baby gradually developed pancytopenia with deranged coagulation profile requiring multiple red cell and platelet transfusions. A trial of steroids Dexamethasone was initiated. Unfortunately, the patient died of multi organ failure before any further treatment could be initiated.

DISCUSSION

The estimated incidence of neonatal LCH (LCH diagnosed within 28 days after birth) in the population-based registry was 1-2/1,000,000 in an Austrian/ German/Swiss/Netherlands based study.[1] Disease in premature infants is rare, with only case reports published; presentation and prognosis vary significantly.[2,3] Disease is classified as single-system (SS-LCH) or multisystem (MS-LCH). In neonates, cutaneous disease is the most common form of SS-LCH and accounts for about 5% of all LCH.MS-LCH, defined by involvement of two or more organ systems. Cutaneous lesions are the predominant presentation in neonates with SS-LCH and MS-LCH.[4] Multisystemic involvement includes lymphadenopathy, hepatosplenomegaly with various degrees of hepatic dysfunction. Gastrointestinal manifestations include bloody diarrhoea often with failure to thrive. Ophthalmological as well as ear manifestations including deafness can be found in upto 1/3rd of the cases. The risk organs as defined in latest literature include liver, spleen and the bone marrow which may cause anemia and thrombocytopenia. CNS involvement is rare and currently has no definitive treatment.

Histopathological examination and immunohistochemical analysis are diagnostic. The main criterion is positivity for CD1a and/or Langerin (CD207). According to latest treatment protocols, treatment for the multisystemic form begins with a six-week course of chemotherapy using vinblastine and prednisone. A second cycle is advised for patients with affected or at-risk organs (such as the liver, spleen, or bone marrow) who show partial or complete response to the first cycle, as well as for those without initial organ involvement but who did not achieve remission. If there are still lesions in at-risk organs after the second cycle or if the disease progresses within six weeks, second-line therapy is recommended. There is no established treatment for patients who do not respond to the initial therapy.[5] Experimental therapies are only recommended for unresponsive disease often in very young patients and includes immunosuppressive therapy with cyclosporine or antithymocyte globulin and possibly imatinib.[6]

CONCLUSION

Multisystemic LCH is a multifaceted disease which has a poor prognosis. Congenital LCH requires a high degree of clinical suspicion to ensure timely diagnosis and initiation of proper treatment protocols. The disease is under investigation for experimental therapies that may improve foetal outcomes, lead to better understanding of disease pathology and overall decrease in mortality and morbidity due to LCH.

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