Severe ocular and risk organ involvement in a newborn with Langerhans Cell Histiocytosis

Alice Brambilla[1], Francesco Pegoraro[1], Maria Luisa Coniglio[1], Roberto Caputo[1], Anna Perrone[1], Anna Maria Buccoliero[1], Claudio Favre[1], and Elena Sieni[1]

[1]Azienda Ospedaliero Universitaria Ospedale Pediatrico Meyer

May 20, 2020

Abstract

Langerhans Cell Histiocytosis (LCH) is a rare proliferative disease of the mononuclear phagocyte system, characterised by tissue infiltration of CD1a+ CD207+ histiocytes. The clinical presentation is variable, ranging from unifocal asymptomatic bone involvement to life-threatening multisystem disease, requiring aggressive therapeutic approaches. Intraocular involvement is uncommon and associated to poor visual and general prognosis. Herein, we report a case of LCH in a newborn with ocular and multisystem risk organ involvement, unresponsive to several lines of chemotherapy. Off-label administration of vemurafenib led to dramatic improvement at systemic level; however, chronic sequelae of ocular involvement resulted in poor visual prognosis.

Hosted file

`Manuscript.doc` available at https://authorea.com/users/324400/articles/452601-severe-ocular-and-risk-organ-involvement-in-a-newborn-with-langerhans-cell-histiocytosis

Hosted file

`Table 1.docx` available at https://authorea.com/users/324400/articles/452601-severe-ocular-and-risk-organ-involvement-in-a-newborn-with-langerhans-cell-histiocytosis