# CONGENITAL NON-LANGERHAN CELL HISTIOCYTOSIS PRESENTING AS BLUEBERRY MUFFIN BABY

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## INTRODUCTION

- *'Blueberry muffin baby'* is a term representing dermal erythropoiesis at birth[1]
- Morphology includes blue-red macules and firm dome-shaped papules[1]
- The most common causes are congenital infections, hemolytic disease of newborn, langerhan cell histiocytosis, neuroblastoma and congenital leukemia[1]
- We report a rare case of congenital non-langerhan cell histiocytosis (NLCH) presenting as blueberry muffin baby

## CASE DESCRIPTION

- A 25 day old newborn girl born at term to healthy Indian parents presented with purpuric rash for the past 10 days
- The neonate had multiple blue-red macules and papules over face, scalp, trunk & upper limbs
- There was no lymphadenopathy or organomegaly and no café-au-lait macules

Fig a Fig b Fig c

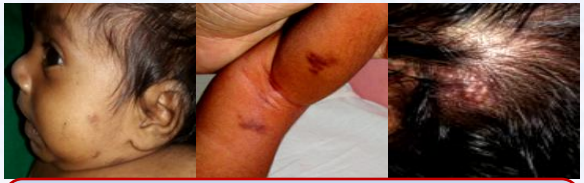
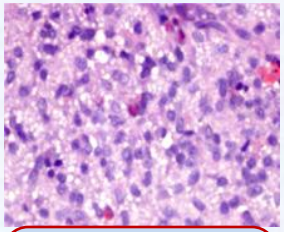
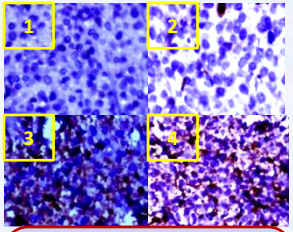


**Fig a & b:** Multiple blue-red non-blanching macules and papules over face (a) and upper limb (b)
**Fig c:** Single red brown lobulated plaque of size 2x2cm over vertex of scalp

Fig d Fig e






**Figd(40x):** Monomorphic infiltrates with vesicular nucleus, vacuolated cytoplasm and few eosinophils

**Fig e1 & e2:** CD1a & S100 IHC stain negative
**Fig e3 & e4:** CD68 & CD14 IHC stains positive

- Hemogram, TORCH serologies, chest X-ray, abdominal and transfontanellar echoes and ophthalmological evaluation were normal
- Hence, we made a diagnosis of congenital NLCH

## DISCUSSION

- Congenital NLCH has been reported to manifest as either juvenile xanthogranuloma (JXG) or benign cephalic histiocytosis (BCH)[2]
- A phenotypic overlap was apparent between these two disorders [2]
- Xanthomatization and touton giant cells were rarely encountered in BCH[2]
- However, in early JXG histology can be indistinguishable from BCH[2]
- Immature lesions may lack xanthomatization and their typical yellow colour[3]

## CONCLUSION

- NLCH should be considered in the differentials of blueberry muffin syndrome
- Absence of typical yellow colour can be misleading

## REFERENCES

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