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CUTIS MARMORATA TELANGIECTATICA CONGENITA – A CASE REPORT

Dermatology

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ABSTRACT

Cutis marmorata telangiectatica congenita otherwise known as Reticular vascular naevus characterized by a bluish purple reticulate pigmentation which may or may not be associated with systemic involvement. Here we report one such case with cardiac involvement in the form of Ventricular septal defect.

KEYWORDS

Cutis marmorata telangiectatica congenita, systemic, cardiac

INTRODUCTION

Cutis marmorata telangiectatica congenita is a vascular malformation of capillaries and veins. It can present either as a localized form predominantly over the extremities in an asymmetrical pattern or as a generalized form. In addition to these, sporadic and familial forms have been described.

CASE REPORT

A 5 year old male child reported to our department with complaints of erythematous pigmentation over the right forearm since birth but otherwise asymptomatic. Patient gave a history of resolution of the lesions with age. Patient gave a positive history of wheeze. No family history of similar complaints. On further investigation, he was diagnosed with a small muscular Ventricular sepal defect of 0.3 cm.

On Dermatological examination, localized erythematous reticulate pigmentation was present over the extensor aspect of right forearm extending from the wrist upto the middle third forearm. [Figure 1] Diascopy was negative. Café au lait macules were present on the lateral aspect of the right shoulder. [Figure 2]

DISCUSSION

Cutis marmorata telangiectatica congenita, described by Von Lohuizen, is a combined vascular malformation present since birth either as a localized or generalized reddish purple telangiectatic reticulate pigmentation. The skin over the reticulate pigmentation may be normal, atrophic or ulcerated.

Various factors are implicated in the pathogenesis like AD inheritance, dermatogens [1]. Congenital anomalies have been found to be associated in 20-70% cases. [2][3]

The most common defects are limb hypoplasia [4], hypertrophy of limbs, aplasia cutis, cleft palate, skeletal defects, cardiovascular defects, dental anomalies. Diagnosis is usually clinical; however screening for the other systemic associations is mandatory to assess the prognosis.

In the absence of systemic involvement, prognosis is usually good, with spontaneous resolution of telangiectatic erythema the norm within 2 years of life.

The closest differential that can be considered is neonatal SLE, which can be differentiated with serological antibody assay.

CONCLUSION

This case has been reported here for its rarity and the association with a ventricular septal defect.

ACKNOWLEDGMENT – None

CONFLICT OF INTEREST – The authors declare that they have no conflict of interest.

LEGEND TO THE IMAGES

Figure 1 – A clinical picture showing erythematous reticulate pigmentation over the forearm

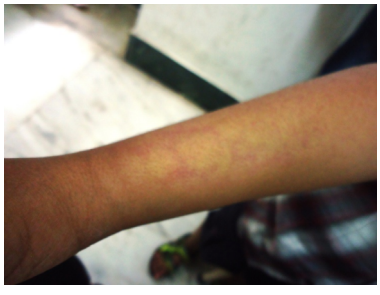
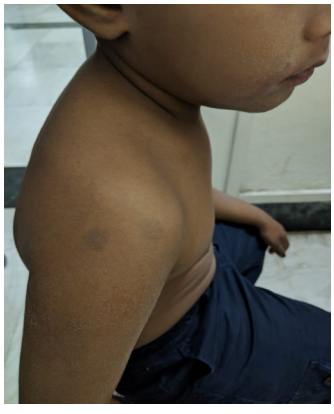


Figure 2 – Clinical picture showing Café au lait macules



REFERENCES

1. Bhargava P, Kuldeep CM, Mathur NK. Cutis marmorata telangiectatica congenita with multiple congenital anomalies: Further clues for a teratogenic cause. Dermatology 1998; 196:368-70.
2. Picascia DD, Esterly NB. Cutis marmorata telangiectatica congenita: Report of 22 cases. J Am Acad Dermatol 1989; 20:1098-1104.
3. South DA, Jacobs AH. Cutis marmorata telangiectatica congenital (congenital generalized phlebectasia). J Pediatr 1978; 93:944-9.
4. Petrozzi JW, Rahn EK, Mofenson H, Greensher J. Cutismarmorata telangiectatica congenita. Arch Dermatol 1970; 101:74-7