Letter to the Editor

Isolated café-au-lait macules: Think of neurofibromatosis type V

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Sir,

Segmental neurofibromatosis (SNF) is a rare form of neurofibromatosis (NF) with a reported prevalence of 0.0014%–0.002%. It is included in Riccardi's classification as type V NF [1].

It is characterized by café-au-lait macules and/or neurofibromas distributed in only one dermatome, less commonly in two or more dermatomes [2]. Roth et al. reclassified SNF into four subtypes: true SNF, localized SNF with deep involvement, hereditary SNF, and bilateral SNF. A limited number of cases of SNF have been reported with systemic involvement, such as visceral neurofibromas, skeletal abnormalities, and renal agenesis [1]. Laser therapy may be performed if an aesthetic demand arises.

Herein, we report a case of SNF in a young boy with no systemic disease.

An eleven-year-old male was sent by his pediatrician for skin spots that appeared at the age of four. The young patient came from a non-consanguineous marriage and had no family history of skin disease. A general physical examination revealed normal parameters, such as weight, size, intelligence, speech, auditory function, and visual acuity. A dermatological examination found unilateral café-au-lait macules with a ranging size of 1 to 5 mm in the right half of the trunk with no crossing of the midline (Figs. 1a and 1b). We thoroughly examined the boy and found no other features of neurofibromatosis, including neurofibromas, neurological deficits, or bone abnormalities. We assured the patient and his parents that this was a benign disease, so the risk of developing any disease-related complications was low. Annual monitoring was started. No therapy was proposed because the patient expressed no aesthetic demands.

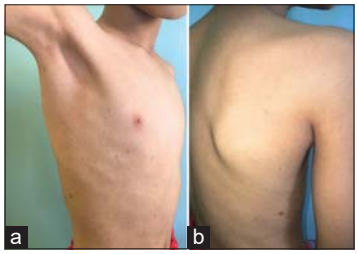

Figure 1a and 1b: Unilateral cafe-au-lait macules of different sizes in the right half of the trunk with no crossing of the midline.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

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