CASE REPORT
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IDIOPATHIC ERUPTIVE MACULAR PIGMENTATION

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Abstract

Idiopathic eruptive macular pigmentation (IEMP) is an rare, rather under reported disease entity of the pediatric age characterized by asymptomatic, brownish hyperpigmented macules involving the neck and trunk with no preceding inflammation or exposure to drug. Here we report this case to increase awareness of this entity among dermatologists, dermatopathologists and pediatricians. A 23 years old healthy female presented with brown-gray to dark, discrete, pruritic macules on the , trunk, neck and limbs (face is spared)of insidious onset. Histopathologic examination showed acanthosis,melanophages, mild perivascular lymphohistiocytic infiltrate in the papillary dermis and papillomatosis. The natural course of the disease is spontaneous remission without treatment, which was not seen in our patient. IEMP is a benign entity with an excellent prognosis as it exhibits spontaneous resolution. It falls into the differential diagnoses of asymptomatic hyperpigmentatory disorders in pediatric population. Awareness of the entity leads to avoidance of unnecessary aggressive damaging treatment.

Key words: Idiopathic eruptive macular pigmentation, papillomatosis

Introduction

Idiopathic eruptive macular pigmentation (IEMP) is a pigmentary disorder of unknown etiology. It was first reported by Degos et al.,[1] in 1978. Knowledge and familiarity of this entity is minimal. Less than 30 cases of IEMP have been reported in the literature so far, reflecting unfamiliarity with the entity.[2]

Case Report

This is a case report of a 23 years healthy girl who presented with pruritic brown to dark lesions over the neck, trunk and proximal extremities of 8 years duration. They appeared spontaneously without any preceding lesions or topical therapy. The lesions started insidiously and gradually progressed over a duration of 3 month.

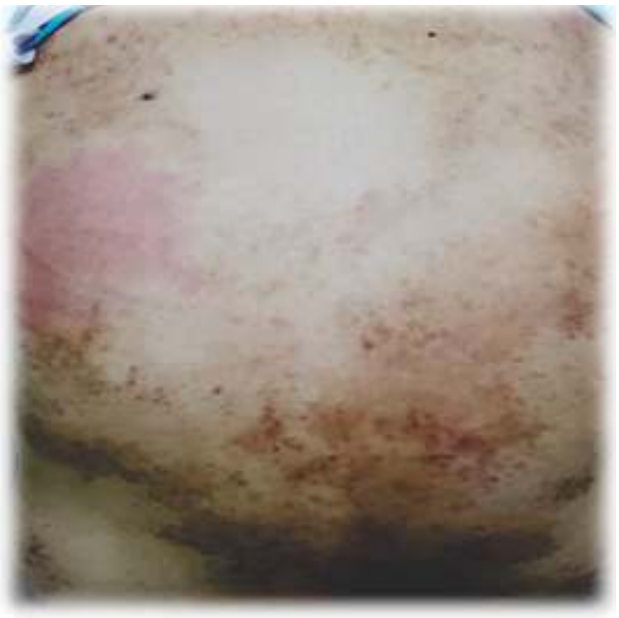

Figure 1 : Hyperpigmented macules present over back

On Cutaneous examination, multiple brownish gray to dark, discrete, round to oval macules ranging from 0.3 to 1 cm in diameter were seen [Figures 1]. On back, there was a large capillary hemangiom, measuring 10x8 cm [Figures 2].The lesions on the lower abdomen had a velvety texture. Face ,Palms and soles were spared. Darier's sign was negative. Other general physical examination were unremarkable. Hematological investigations were normal.

Figure 2 : Large size capillary hemangioma present over back since birth

Biopsy showed moderate irregular acanthosis, papillomatosis [Figure 3], basal layer hyperpigmentation And upper dermis showed sparse superficial lymphohistiocytic infiltrate [Figure 4]. Few melanophages were seen in the papillary dermis. The mast cell number was normal [Figure 5]. The final diagnosis was IEMP. Patient is treated with topical steroid and oral

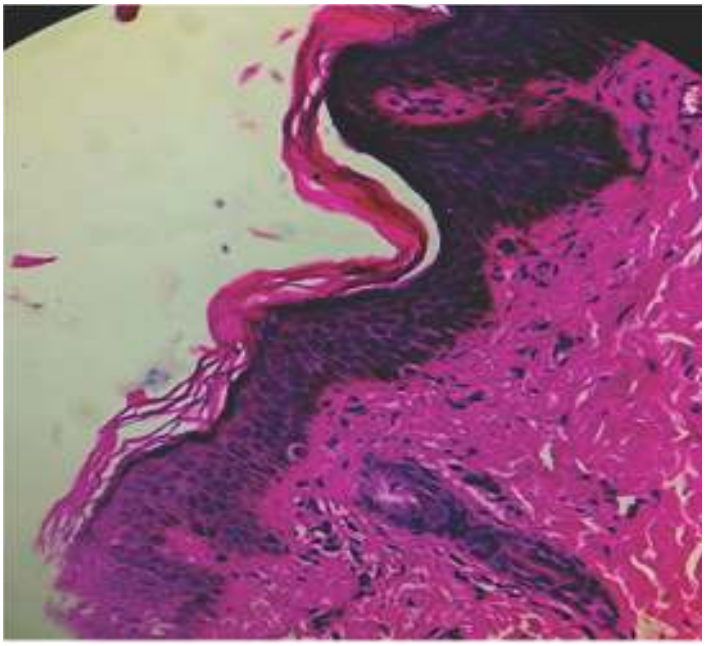

Figure 3 : Histopathology shows acanthosis,papillomatosis and sparse superficial lymphohistiocytic infiltrate in upper dermis. (HxE(4x))

antihistaminics.Patient show excellent response within 15 days . Topical Timolol is also advised to patient for capillary hemangioma which show no satisfactory response.

Discussion

IEMP is a rare and under diagnosed condition. Although it clinically resembles to lichen planus pigmentosus, erythema dyschromicum perstans, fixed drug eruption and mastocytosis. Therefore the only way to differentiate is by histopathological examination. The youngest and oldest case reported in the literature is that of a 1-year-old and a 50-year-old.[3,4] Though most cases have been reported in the young; it has also been reported in a 31-year-old female.[5] A study by Sanz de Galdeano et al.[6] gave the criteria for diagnosis in 1996, namely: (a) Eruption of brownish, non-confluent, asymptomatic macules involving the trunk, neck and proximal extremities in children and adolescents (b) absence of preceding inflammatory lesions (c) no previous drug exposure (d) basal layer hyperpigmentation of the epidermis and prominent dermal melanophages without visible basal layer damage or lichenoid inflammatory infiltrate (e) normal mast cell count. The present case fulfilled all the above mentioned criteria and very few cases of similar nature have been reported [2,7] among Indians.

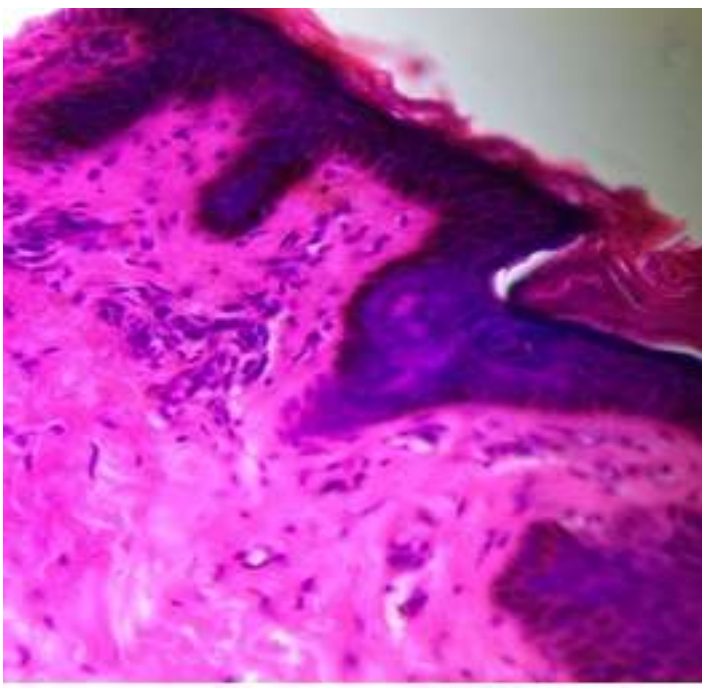

Figure 4 : HXE showing papillomatosis

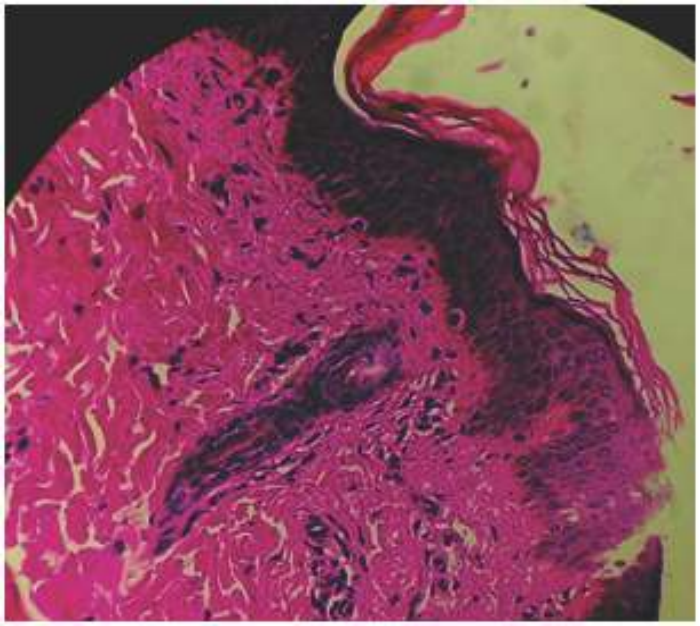

Figure 5 : :HXE showing mast cells.

The histopathological finding,`pigmented papillomatosis` is characteristic feature which was seen in the present case as well. IEMP is self-resolving and has been reported to disappear spontaneously in months to years. The largest series of ten cases have been described by Jang et al.[3]

This case is reported for its rarity , as most cases of IEMP shows spontaneous resolution but this case did not show spontaneous resolution. Most of the cases of IEMP are asymptomatic clinically but this case shows significant itching which shows excellent response with topical steroid and antihistaminics.

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