

Case report

Dermatopathia Pigmentosa Reticularis with Salzmann's nodular degeneration of cornea: A rare association.

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Abstract

Background: Dermatopathia pigmentosa reticularis (DPR) is a very rare autosomal dominant disorder with the diagnostic triad of generalized reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy. **Objective:** To describe the occurrence of Salzmann's nodular degeneration of cornea with moderate dry eye in a patient with Dermatopathia pigmentosa reticularis. **Case:** We present an 11 year old young Indian girl with DPR who had Salzmann's nodular degeneration of cornea with moderate dry eye. She was put on symptomatic treatment and counseled regarding the course of disease, familial nature and avoidance of exposure to sun. **Conclusion:** In a patient of Salzmann's nodular degeneration with generalized reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy the diagnosis of DPR must be kept in mind. A multidisciplinary approach is required for the management of such cases.

Keywords: Dermatopathia pigmentosa reticularis, Salzmann's nodular degeneration, corneal opacity, dry eye, autosomal dominant

Introduction

Dermatopathia pigmentosa reticularis (DPR) is a rare autosomal dominant disorder with the diagnostic triad of generalized reticulate hyperpigmentation, noncicatricial alopecia and onychodystrophy. Other dermatologic findings associated with this triad include adermatoglyphia, hypohidrosis or hyperhidrosis, palmoplantar hyperkeratosis, and acral dorsal nonscarring blisters (Bu TS et al, 1997). We report a case of DPR with bilateral Salzmann's nodular corneal degeneration with moderate dry eye.

Received on: 08/11/12

Accepted on: 26/07/14

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Case report

An 11 year old young girl presented with complaints of hyperpigmentation all over the body, hair fall, and nail changes since 2 years of age. There was a history of gradual, progressive, painless diminution of vision and irritation of both eyes for the past 4 years. The parents did not have any such complaints and she did not have any siblings. The physical examination of patient showed presence of generalized reticulate hyper pigmentation all over the body more pronounced on the extremity, onychodystrophy (figure 1) and non scarring alopecia (figure 2). Ophthalmic examination revealed unaided visual acuity of 6/9 in both eyes which did not improve with refraction. Slit lamp examination of cornea showed discrete elevated, avascular, oval, bluish-white

nodular superficial corneal opacities reaching up to the pupillary margin in both the eyes (figure 3). The corneal findings were consistent with Salzmann's type nodular degeneration. Schirmer test resulted in moderate dry eye. There was no evidence of follicles or scarring on palpebral or bulbar conjunctiva. Rest of the ocular examination was normal. Neurological, cardiovascular, respiratory and abdominal examination was unremarkable. Impression cytology of the conjunctiva revealed squamous metaplasia and decreased density of goblet cells. Blood coagulation studies, blood chemistry analysis and urine analysis were unremarkable. Nail clippings were negative for fungus. Chest x-ray and montoux test was normal. Upper Gastrointestinal endoscopy and CT scan abdomen were normal. Genetic analysis revealed that the disease could be mapped to band 17q. The patient was prescribed sodium carboxy methyl cellulose 1% eye drops thrice a day in both the eyes. She was put on topical retinoic acid for the hyperkeratosis and advised to avoid sun exposure to prevent blister formation. She was explained about the unabating nature of the pigmentation. Genetic counseling of her family was done explaining the autosomal dominant pattern of inheritance.



Figure 1: Photograph of hands showing onychodystrophy of nails and reticulate pigmentation.



Figure 2: Photograph of scalp showing non scarring alopecia.



Figure 3: Photograph of left eye showing Salzmann's nodular degeneration.

Table 1: Differential diagnosis of dermatopathia pigmentosa reticularis.

	Pigmentation	Alopecia	Nail	Palmplantar Keratoderma	Other
Our patient	Generalized reticulate pigmentation since the age of 2 years	Non scarring alopecia	Onychodystrophy With pterygium	Absent	
Dermopathia pigmentosa Reticularis	Generalized reticulate pigmentation	Present	Onychodystrophy With pterygium	Present	Sweating disorders; decreased dermatoglyphics;
Naegeli-Franceschetti-Jadassohn syndrome	Reticulate pigmentation Fading of pigmentation	Absent	Onychodystrophy	Present	enamel hypoplasia; hyperhidrosis; nail dystrophy absence of dermatoglyphics, heat intolerance, punctuate keratosis.

Dyschromatosis universalis hereditaria	Reticulate pigmentation starts on the extremities and involves the whole body	Absent	Pterygium	Absent	small stature, and high tone deafness
Mendes da Costa disease	Generalised, face and limbs	Absent	Present	Present	Blistering early; hypopigmented macule Microcephaly; mental retardation; atrichia; short conical fingers
Dyskeratosis congenita	Can be generalized	Absent	Present	Present	X-linked recessive gene, and leukoplakia and hematologic abnormalities are highly characteristic

Discussion

Dermatopathia pigmentosa reticularis was first described by Hauss and Oberste Lehn (1958). The pigmentation of dermatopathia pigmentosa reticularis occurs at birth or during early childhood (Bu TS et al, 1997). The pigmentation is generalized but most prominent on the trunk and proximal extremities. The features that help to differentiate it from other generalized reticulate disorders (table 1) are the progressive alopecia involving the scalp, eyelashes, and eyebrows and axillae and onychodystrophy of nails. Progressive disease results in loss of nails, with pterygia formation often during the second year of life.

A unique feature unreported in any other reticulate pigmentation disorder is fine punctate spots on the cornea. Amongst the differentials entertained (Table 1) this feature associated with superficial raised bluish white nodule suggestive of Salzmann's nodular corneal degeneration helped clinch the diagnosis. Salzmann's nodular degeneration has been associated with trachoma, vernal keratoconjunctivitis, phlyctenular keratoconjunctivitis, ocular trauma, scarlet fever, and may be a special form of corneal scarring whose pathogenesis is unknown. Dermatopathia pigmentosa reticularis is an uncommon clinical condition with very few case reports in world literature and to our knowledge its occurrence with corneal opacity is reported in very few cases (Gahlen W, 1964; Hauss H et al, 1958;

Van der Lugt L, 1970; Flagel H, 1970). Our patient showed typical features of DPR triad along with corneal features suggestive of Salzmann's nodular corneal degeneration with moderate dry eye. Though there is no definitive treatment for DPR, the patient requires symptomatic management, advice to avoid sun exposure and genetic counseling. Also a close vigil is required for occurrence of gastric carcinoma (Tunca M, 2008).

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Source of support: nil. Conflict of interest: none