Case for diagnosis*
*Caso para diagnóstico**

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HISTORY OF THE DISEASE

A 15-year-old male patient, pheoderma, made reference to one-year long dark, assymptomatic relief lesions on the left hip. The clinical examination showed discretely keratotic confluent papules in plaques, brownish in color and linearly distributed on the left hip. (Figure 1)

Histopathology showed epidermis with mature basaloid cell proliferation forming interwoven beams, besides hyperkeratosis with pseudohorncysts and basal layer hyperpigmentation (Figure 2). There was also a papillary dermis edema, with abundant deposition of mucin, made evident by Alcien blue staining (pH 2.5). Toluidine blue staining revealed metachromasia due to the presence of hyaluronic acid (Figure 3), and orceine staining showed a diminishing of elastic fibers in the papillary dermis.

COMMENTS

The mucinous nevus (MN) is a rare form of primary cutaneous mucinosis.[1] The literature reviewed up until December 2005 allowed for the identification of only nine cases of MN.[1-5] A condition of recent description, with a debatable nosologic situation between the connective tissue nevus and the epider-

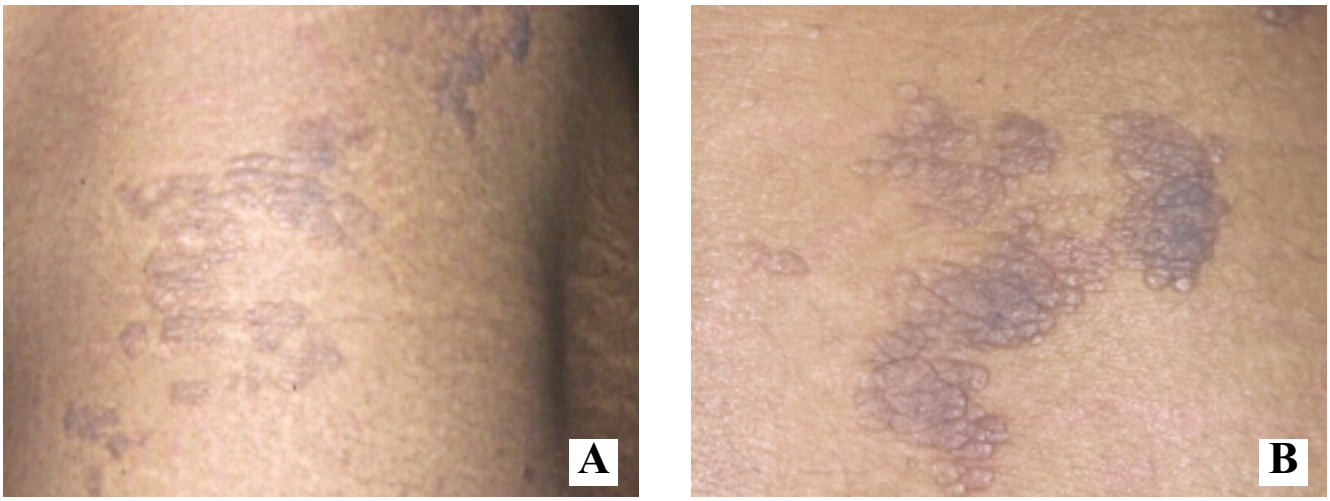

Figure 1: A – Brownish plaques in linear distribution on hip; B – Detail of confluent papular lesions in plaques

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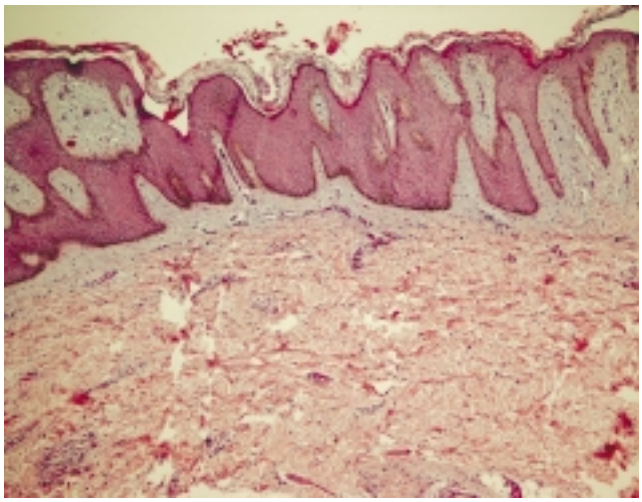

Figure 2: Hyperplasia of the epidermis, hyperpigmentation of the basal layer and diffuse deposition of homogeneous material on the papillary and upper dermis (HE, 20x)

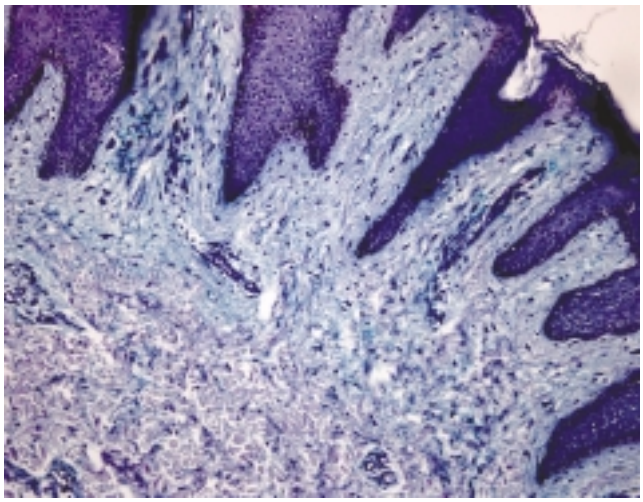

Figure 3: Metachromasia of the papillary and upper dermis (toluidine blue, 40x)

mal nevus, MN was first described in 1993 by Bellón et al., who proposed the name by virtue of the nevoid character and distinctive histological pattern with mucin deposition on the papillary dermis.[2]

The nevoid aspect of the lesions and the early onset suggest a hamartoma, which can be congenital ou acquired.[1] Clinically it appears as multiple papules or brownish papulous plaques present at birth or which appear during adolescence, sometimes tending towards verrucous growth of unilateral distribution, either linear or zosteriform.[1-5] It affects more often the lower area of the trunk.[2-4]

Histopathological findings are characteristic. There is a homogeneous and diffuse deposition of mucin in the upper dermis, totally filling the dermal papillae. Because it is made up of acid glycosaminoglycans, this material presents a positive reaction to staining with Alcian blue (pH 2.5) but negative when pH is 0.5. The mucin deposited in the dermis is essentially composed of hyaluronic acid.[1] Rongioletti & Rebora stress the absence of collagen and elastic fibers in the mucinous deposition area.[1] However, two cases presented an increase of collagen fibers and one of them also vascular proliferation, thus suggesting the possibility of increase of other dermal components in MN.[3] In the presented case, there was a decrease of elastic fibers in the papillary dermis. Besides the dermal deposition of mucin, practically every case presented epidermal changes, including hyperkeratosis, acanthosis, and papilomatosis, which led Rongioletti & Rebora to consider MN as a form of hamartoma which would combine the features of the epidermal nevus with those of the connective tissue nevus of the proteoglycan type.[1]

Due to the nevoid aspect, MN can be clinically confounded with the epidermal nevus, connective tissue nevus, nevus lipomatosus superficialis, and other surface hamartomas.[3,4] It must also be differentiated from other types of cutaneous mucinosis,[1] such as the cutaneous mucinosis of infancy,[4] the cutaneous focal mucinosis and the papular mucinosis, besides other cutaneous conditions that may undergo mucinous degeneration, such as the mucinous eccrine nevus.[1] In papular mucinosis, the mucin deposit is seen in the reticular dermis and around dermal vessels.[1] In cutaneous mucinosis of infancy, the mucin deposit is focal in the papillary dermis whereas in the cutaneous focal mucinosis there is also an increased number of fibroblasts in the dermis.[1]

Whenever feasible, the main therapeutic approach is surgical, whether by total excision of the affected area, used in the two cases described by Lim et al.,[5] or by shaving, which was succesfully used in the case reported by Yokogawa et al.[5] In the case presented here, the surgical approach was not recommended due to the extension and location of the lesions. For this reason, the adopted conduct was one of expectation. ❑

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Abstract: A case of mucinous nevus is reported in a male teenager who presented assymptomatic brown papules confluent in linear plaques in the left hip since the age of 14 years old. Further on mucin deposition in superficial dermis, the histopathology showed reduction of elastic fibers in the papillary dermis, hiperkeratosis, acanthosis and papillomatosis.
Keywords: Glycosaminoglycans; Mucinoses; Nevus

Resumo: *É relatado um caso de nevo mucinoso em adolescente do sexo masculino que apresentava lesões papulosas acastanhadas assintomáticas, confluentes em placas de distribuição linear no quadril esquerdo, surgidas aos 14 anos de idade. Além de deposição de mucina na derme superficial, a histopatologia demonstrava diminuição das fibras elásticas na derme papilar, hiperceratose, acantose e papilomatose.*
Palavras-chave: Glicosaminoglicanos; Mucinoses; Nevo

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