IMAGING CASES

Large brown macula in a newborn

Mácula gigante num recém-nascido

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The authors report the case of a term newborn boy born at 40 weeks postmenstrual age (maternal age 31 years) from an uneventful nonconsanguineous pregnancy monitored in primary health care. Blood group was A Rh-, both parents were healthy, and group B streptococcus screening test was negative. Prenatal ultrasound and serologies were unremarkable. The boy was born by vaginal delivery with a weight of 3530 g, height of 49 cm, head circumference of 33 cm, Apgar score of 9/10/10, good adaptation to extrauterine life, and no oxygen requirements.

Initial physical examination revealed a homogeneous, light brown, hyperpigmented macula more than fifteen cm in diameter on the right upper limb and thoracic region (Figures 1-3). Dermoscopy was performed, revealing a typical brown globular pattern with no irregular structures or visible atypia.

What is your diagnosis?



Figures 1-3. Newborn on the first day of life with a homogeneous, brown, hyperpigmented macula more than 15 cm in diameter on the right upper limb and thoracic region.

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DIAGNOSIS

Giant congenital melanocytic nevus (CMN) - Size at birth >15 cm, with predicted adult size >40 cm.

DISCUSSION

After diagnosis, the newborn was discharged with indication for Neonatology and Dermatology follow-up. On reassessment, he showed adequate psychomotor growth and development, an unremarkable neurological examination, and no areas of concern on routine dermoscopy. Subsequent evaluation with cranioencephalic magnetic resonance imaging (MRI) at 14 months of age was normal.

CMN is a benign proliferation of neural crest-derived melanocytes that form theca or nests at the dermal-epidermal junction and/ or in the dermis.⁽¹⁾ Brown in color, it may be present from birth or appear during the first two years of life at any body surface area.^(1,2) Based on adult size projection, CMN are classified as small (<1.5 cm), medium (1.5-20 cm), large (20-40 cm), giant (>40 cm), or multiple. ⁽³⁾ Compared to other CMN, giant CMN is a rare entity, with an incidence of 1:20,000 newborns, and has a higher and earlier ris for melanoma (6-20%).^(1,2,4,5) Melanomas arising from congenital nev are rarer than spitzoid melanomas, but account for most melanoma related deaths in childhood.⁽⁵⁾

Management of CMN should be based on age, nevus size, and other factors such as psychosocial impact, association with vascula malformations, limb atrophy and asymmetry, neurocutaneou melanosis, and characteristic facial morphology.^(1,2,4,6) Given the potential for malignancy, giant CMN require clinical surveillance with neurodevelopmental follow-up, regular dermoscopy, and early surgical intervention (excision or biopsy) of suspicious lesion for timely diagnosis of melanoma.^(2,4-6) Surveillance with digita dermoscopy has optimized early diagnosis of melanoma and reduce the number of unnecessary excisions. Thus, in lesions considered atypical but with low suspicion of melanoma, dermoscopi surveillance may be preferred to surgery.^(2,4) A spectrum of centra nervous system abnormalities has been described in associatio with congenital melanocytic naevi.⁽⁶⁾ In this context, cranioencephali MRI should be performed for early detection of melanin depositior especially in the temporal lobes, cerebellum, pons, and/or medulla in neurologically asymptomatic patients with giant CMN.^(6,7) | cranioencephalic MRI shows no changes, it should be repeated onl if suspicious symptoms arise.^(5,7)

ABSTRACT

The authors report the case of a male term newborn born after an uneventful monitored pregnancy. On physical examination, he presented with a light brown hyperpigmented homogeneous macule >15 cm in size compatible with a giant congenital melanocytic nevus (CMN). The boy was discharged with indication for Neonatology and Dermatology follow-up. Since then, he has shown adequate psychomotor growth and development and unremarkable neurologic examination and cranioencephalic magnetic resonance imaging.

CMN is a benign proliferation of melanocytes forming theca/nests in the dermal-epidermal junction and/or dermis. It may be present at birth or develop during the first two years of life in any body area. CMN can be classified as small, medium, large, giant, or multiple. Giant CMN is rare and is a risk factor for melanoma.

Management of CMN should be based on age, nevus size, and other factors. Because of the potential for malignancy, giant CMN requires clinical surveillance with neurodevelopmental monitoring and regular dermoscopy, as well as early surgical management of suspicious lesions.

Keywords: congenital melanocytic nevus; dermoscopy; newborn; risk for malignant transformation; skin lesion

RESUMO

É descrito o caso de um recém-nascido de termo, do sexo masculino, fruto de gestação vigiada sem intercorrências. Ao exame objetivo, o rapaz apresentou uma mácula hiperpigmentada, de coloração castanho claro, homogénea, com mais de 15 cm, compatível com nevo melanocítico congénito (NMC) gigante, tendo sido referenciado para a consulta de Neonatologia e Dermatologia. Desde então, tem apresentado crescimento e desenvolvimento psicomotor adequados e exame neurológico e ressonância magnética cranioencefálica sem alterações.

O NMC é uma proliferação benigna de melanócitos, de coloração castanha, que forma tecas/ninhos na junção dermoepidérmica e/ou derme. Pode estar presente ao nascimento ou surgir nos primeiros dois anos de vida em qualquer localização. Os NMCs podem ser classificados em pequenos, médios, grandes, gigantes ou múltiplos. O NMC gigante é raro e representa um fator de risco para melanoma. A abordagem do NMC depende da idade e do tamanho do nevo, entre outros fatores. Dado o potencial de malignidade, o NMC gigante requer vigilância clínica com acompanhamento do neurodesenvolvimento e dermatoscopia regular e abordagem cirúrgica precoce perante lesões suspeitas.

Palavras-chave: dermatoscopia; lesão cutânea; nevo melanocítico congénito; recém-nascido; risco de transformação maligna

AUTHORSHIP

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