Large Segmental Nevus Spilus Associated With Congenital Spondylolysis

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Received date: Dec 18, 2014, Accepted date: Mar 26, 2015, Published date: Mar 30, 2015

Abstract

Nevus spilus or speckled lentiginous nevus is a mixed nevus and presents as a light-brown macule with macular and/or papular-nodular lesions scattered across it. When lateralized, they can be associated with other anomalies, particularly neurological and musculoskeletal becoming the nevus spilus syndrome.

We report a 16-year-old boy with a giant segmental nevus spilus associated with congenital spondylolysis.

Keywords: Nevus spilus; Congenital spondylolysis

Introduction

Nevus spilus or speckled lentiginous nevus is a mixed nevus (“nevus over nevus”) and presents as a light-brown macule (melanotic nevus) with macular and/or papular-nodular lesions scattered across it (melanocytic nevus, often: lentigos, compound nevus cells, junctional nevus and blue nevi, alone or mixed) [1,2]. Two types can be described: a) macular, with regular distribution of flat melanocytic lesions histologically related to junctional nevi or “jentigo”, and b) papular or lentiginous, with an irregular raised melanocytic nevus distribution, histopathologically featured by intradermal or compound melanocytic nevi [3]. Dysplastic nevi can also be found [4]. Although nevus spilus can be multiple (in a small number), they are usually single and not related to another pathology. Nevertheless nevus spilus can be sometimes associated with other anomalies [5-8]. Nevus spilus has also been reported as a precursor of cutaneous melanoma [9,10].

Clinical Case

We report a 16 year old male born with a lentiginous papular nevus spilus over almost the entire right lower limb (Figures 1 and 2).

Figure 1: Lentiginous papular nevus spilus over the right buttock and back side of the thigh.

The nevus had a bigger papular lesion (0.5 cm x 1cm) in the right buttock. Histopathological examination revealed basal hypermelanosis and melanocyte nests in crests (Figure 3). Vertebral anomalies were found at the screening tests. The lumbosacral radiography at maximum flexion showed misalignment in L4-L5 and the computed tomography (CT) scan confirmed the right spondylolysis in L4 (Figure 4). The patient had no symptoms or physical examination of spondylolysis and denied a history of mechanical factors that could trigger it.
Discussion

Nevus spilus typically presents as a small isolated lesion, although large segmental lesions can rarely be found (lentiginous zosteriform nevus) [11]. It is occasionally associated with complex birth defects such as phacomatosis pigmentovascularis in the macular type, or phacomatosis pigmentokeratotica or speckle lentiginous nevus syndrome in the papular type [12].

Congenital type spondylolysis is a congenital bone deficiency development (lumbar vertebral, usually L4 and L5), which lasts lifelong. It has no symptoms, a strong hereditary base and is considered a autosomal dominant condition [13]. There is no treatment, but it needs to be controlled due to the risk of vertebral displacement (spondylolisthesis) [14].

In conclusion, large segmental nevus spilus lesions are very infrequent. When lateralized, they can be associated with other anomalies, particularly neurological and musculoskeletal becoming the nevus spilus syndrome. Approximately 20 cases have been reported. We report a particular association with spondylolysis.

References