Giant congenital melanocytic nevus with Becker's nevus-like presentation

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ABSTRACT

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Congenital melanocytic nevus (CMN) is present at birth, clinically presents as a round brownish lesion with well-defined borders and hypertrichosis. The surface of the nevus may be slightly raised, papular, roughed, warty or cerebriform[1,2]. CMN is often composed of complex three histological patterns that include nevus cell type, neuroid type, and blue nevus cell type. In our case patient presented with hypertrichosis, flat hyperpigmented patches with an irregular border. From this clinical presentation, the differential diagnosis were Becker's nevus, pigmented neurofibroma and congenital melanocytic nevus which can mimicking each other. Skin biopsy was showed tyrosinase-positive spindle cells. In this case, we conclude the diagnosis as congenital melanocytic nevus with neuroid type.

Key words: giant congenital melanocytic nevus, pigmented neurofibroma, Becker's nevus

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Congenital melanocytic nevus (CMN) is present at birth, but some authors defined as CMN that develop from birth until two years (tardive congenital nevi). Late congenital melanocytic nevus may represent insufficient initial production of melanin or small size of nevus, make it difficult to early identify[1-4]. Giant congenital melanocytic nevus (GCMN) is generally defined as a congenital melanocytic lesion that will reach, at least 20cm in adult life, but sometimes refer to more than 40 cm in size[5-8]. About 1% of live birth present with a CMN. The incidence of GCMN is estimated to be from 1:20,000 to 1:500,000 depending on the type of GCMN and references[9,10].

Case Report

A 24-year-old man, presented with localized brownish patches on the left face since he was 3 months old. The lesions gradually enlarged and lesions developed hypertrichosis when he was 15 years old. On physical examination, there were two large localized well-defined irregular margin brownish patches with satellite macules on the left temple and cheek with hypertrichosis (Fig. 1, 2). No clinical findings of café au lait macules, axillary or inguinal freckling, abnormal of a musculoskeletal organ. Ophthalmologic and neurologic examination are within normal limits.

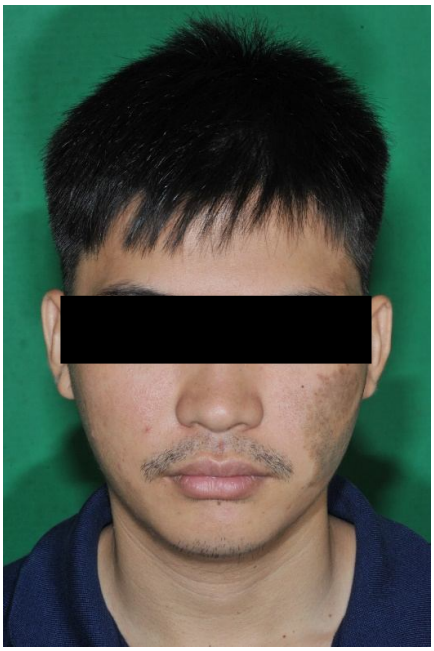

Figure 1 two well defined irregular border hyperpigmented patch on left face with hypertrichosis. Symmetrical of face

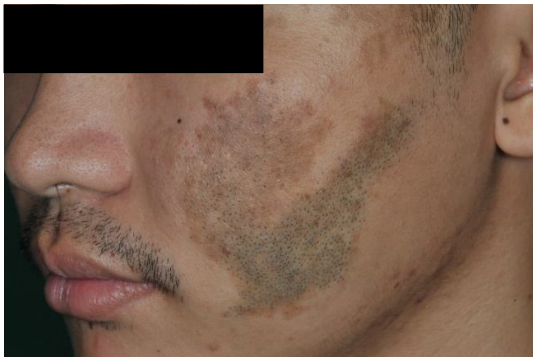

Figure 2 localized well defined irregular border hyperpigmented patches with hypertrichosis and some satellite lesions

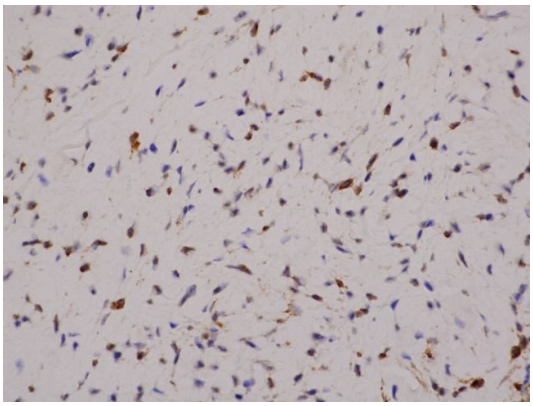

Figure 3 Infiltrative of spindled cell with some pigmented in deep dermis to subcutaneous fat

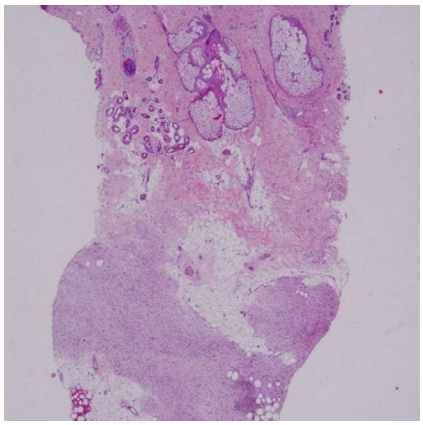

Figure 4 Immunohistochemical study demonstrates immunoreactivity of spindle cell lesion to tyrosinase.

Histopathology was performed and demonstrated epidermal hyperplasia. The dermis showed increased spindle-cell component in deep dermis extending into subcutaneous fat, with some pigmentation (Fig. 3). Immunohistochemical study was done, tyrosinase (Fig. 4) and S100 were strongly positive on spindle-cell component. Smooth muscle actin, neuro-specific enolase, and neurofibromin showed negative expressivity. The patient was diagnosed with congenital melanocytic nevus.

Discussion

CMN clinically presents as a round or oval brownish lesion with well-defined borders and hypertrichosis. The surface of the nevus may be slightly raised, papular, rough, warty or cerebriform[14,15]. The lesion is usually darkening during the first few years of life, some lesions may become lighter or appear as satellite lesions[12].

CMN was originated from neural crest stem cells and migrated to the skin. The larger and deeper-located melanocytes represent the earlier migratory stage of melanoblast from the neural crest, like in GCMN[11]. CMN and acquired melanocytic nevus may show similar histologic findings. The melanocytic cell has many characteristics depends on the site, such as junctional nest has round, ovoid or fusiform shapes and are arranged in a cohesive nest. In the superficial dermis, the cell has epithelioid characteristic and contains amorphous cytoplasm with frequent granular melanin. In deeper reticular dermis, there is diminished

content of cytoplasm, resemble lymphocytic or spindled configuration, similar to fibroblast or Schwann cell[12].

GCMN could presents as three complex histological patterns: nevus cell type, neuroid type, and blue nevus cell type.

Neuroid type is a spindled cell in the dermis that can extend deeply to subcutaneous fat. Three types of neuroid pattern that resembling Wagner-Meissner corpuscles, Verocay body, and sheet of schwannian cell may present around the nerves and blood vessels in columnar or tubular appearance[13]. The diversity of histological elements could be explained that these lesions originate from pluripotent stem cells, which can differentiate into multiple cell types[11].

In our patient, the clinical presentation was hyperpigmented patches, which are an unusual presentation in a giant congenital melanocytic nevus. The differential diagnosis at the time were Becker's nevus and pigmented neurofibroma. Becker's nevus which usually presents at the second or third decade of life, located on the shoulder, upper back, and lateral arm but rarely present on the face**,** hyperpigmented patches with broken or irregular border, and hypertrichosis[12]. Pigmented neurofibroma may be presented as a subcutaneous type with hypertrichosis but the lesion usually presents with firm or soft consistency. Histopathologic of pigmented neurofibroma could also be positively staining with S100 and tyrosinase[16] as in our case but with less expressivity.

Our patient presented with clinically mimicking Becker's nevus and pigmented neurofibroma, but clinical onset and histopathology supported a deep neuroid-type melanocytic neoplasm showing S100 and tyrosinase positivity and negative neurofibromin staining. Histological findings confirmed the diagnosis of congenital melanocytic nevus.

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