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Case Report

Segmental Neurofibromatosis: Two Moroccan Cases

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Abstract

Segmental neurofibromatosis (SN) or type V Neurofibromatosis is a genodermatos most often characterized by the presence of neurofibromas, more rarely café au lait spots, and sometimes lentigines limited to a limited body region. We report two Moroccan cases of true Segmental Neurofibromatosis with only pigmented lesions.

Keywords: Segmental Neurofibromatosis; Type V Neurofibromatosis; Café au lait spots; Neurofibromas; Lentigines

Introduction

Segmental neurofibromatosis (SN) or type V Neurofibromatosis is a genodermatos most often characterized by the presence of neurofibromas, more rarely café au lait spots, and sometimes lentigines limited to a limited body region, or more rarely on several bilateral segments. It is exceptional, with an estimated prevalence between 0.0014-0.002%. We report two Moroccan cases of SN.

Clinical case

Observation N 1: A 10-year-old boy, with no family history of Neurofibromatosis (NF), who has had numerous café-au-lait spots and lentigines at the left lumbar region since the age of 2. No ophthalmologic and neurological symptoms of neurofibromatosis were diagnosed. The diagnosis of segmental neurofibromatosis (NF-5) has therefore been established.

Observation N2: A 35-year-old woman with no concept of consanguinity, followed in neurology for a multiple sclerosis under treatment presented with a 25-year history of coffee-milk spots and lentigines at the right lumbar region.

Comments: The term segmental neurofibromatosis was introduced by Miller and Sparkes in 1977 [1-2]. Segmental NF1 is caused by somatic mosaicism due to a postzygotic mutation in the NF1 gene. This results in some cells containing two normal NF1 genes and other cells having a mutation in one copy of the NF1 gene [3]. Due to the multiplicity of clinical manifestations of NF, Riccardi ranked NF in eight categories in 1982. Type V was reserved for the NS [4-2]. In 1987, [5] Have classified the NS in four variants: true segmental, Localized with deep evolution, hereditary and bilateral. Both of our patients had a true NS.

The most common clinical manifestation of NS is: neurofibromas, and less frequently, coffee-au-lait spots.Clinically, patients can be divided into four groups: NS with only pigmented lesions, NS with only neurofibromas, NS with pigmented lesions and neurofibromas, and NS with isolated plexiform neurofibromas [1,2,4]. Our patients have only pigmented lesions. Systemic involvement in NS is rare, except in patients with plexiform neurofibromas. NF I and NS may be associated with malignant tumors such as peripheral nerve sheath tumor, melanoma, breast cancer, colon cancer, stomach cancer, lung cancer and Hodgkin lymphoma [6] (Figures 1 & 2).

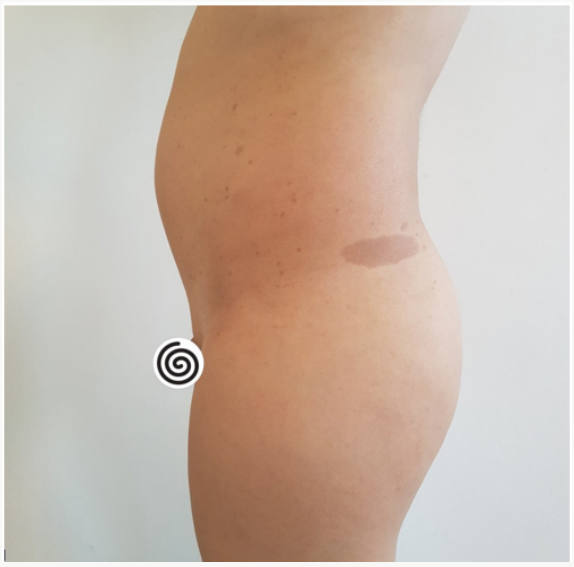

Figure 1: Patient 1 : Multiple lentigines and café au lait spots on the left lumbar region.

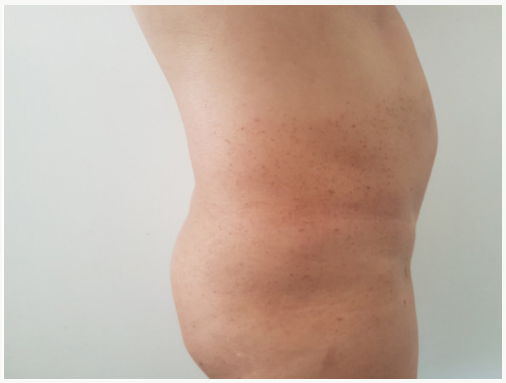

Figure 1: Patient 2: Multiple lentigines and café au lait spots on the right lumbar region.

Conclusion

We present the first two cases of Moroccan SN. The risk of association with malignant tumors or with systemic involvement is not confirmed, so patients presenting SN must have a long-term medical follow-up.

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