

Short Case Report

Two white sponge nevus in a single family: case report

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Abstract – Introduction: White Sponge Nevus (WSN) is a leukokeratosis characterized by white lesions of the oral mucosa. These lesions are bilateral, thickened and raised compared to the adjacent mucosa. Their aspect are folded and spongy. It is a benign disorder with asymptomatic lesions which often appear during the childhood or the adolescence. The interest of this case report is that the diagnostic of WSN had been established directly, thanks to the presence of the patient's father. **Observation:** A twelve years old patient was examined in the unity of the oral mucosa pathology and oro-facials pain, of the oral surgery service of Bordeaux hospital (CHU de Bordeaux, France). He presented typical WSN lesions. His father was examined and presented the same lesions. **Discussion:** Diagnostic of WSN is mainly a clinical examination. There are a lot of differential diagnosis, and leukoplakia is the principal. In case of doubt about the diagnostic, a histological examination can be done. Nowadays, there is no consensus about the therapeutic. But the lesions are mainly asymptomatics, so any treatment has to be planned. **Conclusion:** An early WSN diagnosis avoids to patients a non adapted treatment and reassures young patients and their parents.

Observation

A twelve years old patient, only child, was addressed by his dentist in the unity of the oral mucosa pathologies and oro-facials pains, in the oral surgery service of Bordeaux hospital (CHU de Bordeaux, France). The aim of the consultation was the presence of white lesions on the buccal mucosa. The medical history of the patient revealed an atopic context with asthma and peanut allergy. The patient did not feel any pain nor discomfort. The clinical examination revealed bilateral and symmetrical white lesions, painless, localized on the mucosa in the interior of the cheek. Lesions were slightly swollen (Fig. 1). Not any other extra-oral lesions had been found.

During the appointment, the patient's father was present and the interrogation revealed that he had in the past a diagnosis of oral leukoplakia. The examination of the patient's father had shown the same kind of lesions, and same localization on the oral mucosa than his son. With the typical aspect of the lesions and the hereditary context, diagnosis of WSN has been straightaway affirmed. Complementary examinations were not necessary. Not any treatment was suggested because patient did not feel any symptom. An information

about chronicity nature of these lesions and their good prognosis was given. A regular follow-up was not organized, justified by the benign character of these WSN lesions.

Commentary

This case report shows and reminds that diagnosis of WSN is only clinical, because it is based on the clinical examination of patient's lesions and his family. The clinical aspect of these lesions is explained by a mutation in keratin gene. Keratin is one of the most important protein which compose intermediate filament (IF), fundamental component of the cellular cytoskeleton. There are 54 genes encoding functional keratins. Keratin 4 and keratin 13 are found in the epithelium of the oral, nasal, oesophageal and anogenital mucosa. A patient affected by WSN has mutation on his gene encoding keratin 4 (KRT4) and 13 (KRT13). The mutation is located in exon 1 of KRT4 and KRT13 [1]. In this way, synthesis of intermediate filament is disrupted. The mutation in KRT 4 seems to be the most frequent mutation compared to the mutation in KRT 13 [2].

To confirm the diagnosis of WSN, Benoit and al took blood samples from their patients, and made a genetic analysis [3]. Nevertheless, Cai W and al showed that the diagnosis was first clinical, with usually a familial context of WSN which help to confirm this diagnosis, just as the present case [1].

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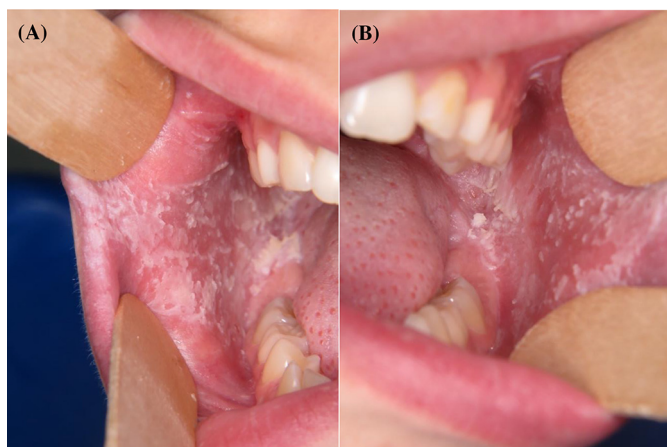


Fig. 1. Clinical aspects of White Sponge Nevus. White and spongy plaques are observed bilaterally on the oral mucosa of the right and left cheek.

To complete the interrogation and the clinical examination, histological examination is occasionally performed, and confirm the diagnosis of WSN. Histopathological findings are a thickened and vacuolization in spinous layer, a hyperkeratosis and an acanthosis. These characteristics explain the white aspect of the WSN lesions. This complementary examination help to do the differential diagnosis with all of the others white lesions as lichen planus or leukoplakia for example. However, it is possible to affirm a positive diagnosis of WSN with simply the clinical examination and the hereditary familial context [2]. In this way, an useless biopsy is avoided and the management of the patient is optimized. In the present case, none of complementary examination was necessary because the diagnosis was only clinical, thanks to the examination of the patient's father.

There are a lot of differential diagnosis: leukoplakia, oral lichen planus, candidiasis, chemical burns, syphilis, trauma, cheek bite, tobacco and betel nut included keratosis [2]. In this case, the patient's father had a wrong diagnosis of leukoplakia, because the white aspect of his mucosa may evoke a leukoplakia.

For children, differential diagnosis are candidiasis, infection by human papilloma virus or dyskeratosis congenita [3]. However, typical aspects of bilateral and symmetrical white

lesions, with spongy aspect, the absence of pain, and the hereditary context allow to confirm positive diagnosis of WSN and exclude all of these differential diagnosis.

About the treatment, many locals and systemics treatment had been investigated to reduce the appearance of the lesions and the altered mucosal texture. McDonagh and al described the efficacy of tetracycline on four patients with WSN lesions [4]. Several cases had been treated with tetracycline mouth rinse, and there was an improvement of the symptoms. Satriano and al showed a regression of white plaques after eight days with chlorhexidine mouthwash 0.12%. But one month after of treatment cessation, plaques reappeared [5]. The mechanism of action of these treatments stay misunderstood. Several authors thought that the clinical amelioration may be explained by the antimicrobial effect of the antibiotics molecules on the mucosa, so microorganism proliferation is reduced and consequently the aspect of WSN lesions [3–5].

Currently, a therapeutic consensus about this disorder doesn't exist. Besides, with absence of symptom or patient complaint, establishment of a treatment is not justified.

About the evolution, it is a chronic and benign disorder.

It is essential that oral surgeons confirm promptly the clinical diagnosis of white sponge nevus. It avoids realization of useless biopsy, prescription of inadequate treatments, and allow to reassures young patients and their parents about the benign character of the lesions.

References

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