Theranostics of
Brain, Spine & Neural Disorders
ISSN: 2641-8096

Juniper Publishers
key to the Researchers

Mini Review
Volume 4 Issue 3 - January 2022
DOI: 10.19080/JOJS.2022.04.555636

Theranostics Brain,Spine & Neural Disord

The Pallid Plat-White Sponge Nevus

Anubha Bajaj*

Consultant Histopatholog, Panjab University, India

Submission: December 20, 2021; **Published:** January 10, 2022

***Corresponding author:** Anubha Bajaj, Consultant Histopatholog, Panjab University, India

Abbreviations: CK: Cyto Keratin; PAS: Periodic Acid Schiff's

Preface

White sponge nevus is an exceptional, hereditary genodermatosis which characteristically depicts asymptomatic, spongy, white plaques usually confined to the oral mucosa. The mucosal disorder exhibits an autosomal dominant mode of disease transmission. White sponge nevus was initially scripted by Hyde in 1909 and Cannon designated the terminology of "white sponge nevus" in 1935 [1,2] The condition is additionally designated as Cannon disease, familial white folded hypertrophy of mucous membranes, hereditary leukokeratosis, white gingivostomatitis or exfoliative leukoedema. White sponge nevus can be appropriately discerned upon distinctive clinical appearance and accompanying clinical history.

Disease Characteristics

White sponge nevus may be congenital or emerge during early childhood or adolescence [3,4]. White sponge nevus predominantly implicates the oral mucosa although sites such as nasal cavity, oesophagus, rectum or vaginal mucosa may be incriminated. Intraoral sites such as the tongue, labial mucosa, soft palate, alveolar mucosa and floor of mouth may exceptionally exhibit lesions of white sponge nevus besides extra-oral sites as the nasal, oesophageal, laryngeal, rectal or genital mucosa [3,4]. The condition is engendered on account of defective keratinization of oral mucosal epithelium with point mutations within genes encoding for cytokeratin (CK) 4 and cytokeratin 13, specifically configured within spinous cell layer of oral mucosal epithelium [3,4]. Generally, cytokeratin 4 and cytokeratin 13 molecules may be modified although cytokeratin 4 gene may remain unaffected. A contemporary mutation affecting cytokeratin 4 (CK 4) gene with amino acid insertion within IA alpha helical domain of cytokeratin CK 4 may engender the condition [3,4]. Deteriorated stability of keratin filament may ensue due to mutation within 2B domain of cytokeratin 4 gene. Also, point mutation within cytokeratin 13 gene may engender aberrant degradation of cytokeratin 13 protein, possibly associated with anomalous ubiquitination [3,4]. Genomic mutation of cytokeratin 4 or cytokeratin 13 genes is observed, which encode mucosa-specific keratin intermediate filament with proteins keratin 4 and keratin 13 and are crucial to assembly of keratin filaments. Sporadic instances of white sponge nevus exhibit keratin mutation in around ~20% instances [3,4]. The genetic disorder with autosomal dominant mode of disease transmission occurs in around one in 200,000 individuals. Frequently, white sponge nevus exhibits variable expression and irregular penetrance [3,4]. Lesions of white sponge nevus may be congenital, appear during early childhood or adolescence. A specific gender predilection is absent [3,4].

Clinical Elucidation

Generally, lesions may be asymptomatic or associated with pain, dental caries or periapical dental radiolucency and manifest as periapical inflammatory disease. Episodic oral mucosal burning upon ingestion of acidic or spicy food may be observed [3,4]. Diffuse, soft, thick, whitish plaques with a corrugated superficial surface are disseminated upon bilateral buccal mucosa, dorsum of tongue, inferior labial mucosa, hard palate, soft palate, retro-commissural area or retro-molar area [3,4]. Oral lesions of white sponge nevus manifest as white or grey, thick, diffuse plaques with multiple furrows and a spongy texture. Generally, oral lesions are located upon buccal, labial or gingival mucosa and floor of the mouth [3,4]. Majority of lesions are bilateral, symmetrical and depict thick, whitish, corrugated or velvety, diffuse plaques. Upon examination, symmetric, irregular, white plaques with a well-defined perimeter appear disseminated upon the buccal mucosa. Erythema is absent. Plaques are usually resistant to abrasive eradication with a blade [5,6]. Commonly, bilateral, symmetrical oral mucosal lesions emerge as thickened, white, velvety, diffuse plaques. Implicated buccal mucosa appears folded with a soft or spongy texture [5,6]. Generally, the incriminated, folded buccal mucosa with a soft or spongy texture demonstrates a distinctive

white, opalescent discoloration [5,6]. Associated ragged, white, non haemorrhagic zones may be expunged with gentle rubbing [5,6]. Clinically manifested plaques may exhibit altered distribution of lesions, magnitude and variation in incriminated zones over a period of time. Infrequently, intraoral white sponge nevus may appear upon the tongue, labial mucosa, soft palate, alveolar mucosa or floor of the mouth. Malignant metamorphosis of white sponge nevus is an exceptional phenomenon although occurrence of oral squamous cell carcinoma within white sponge nevus is documented. Malignant transformation may be induced by chronic, persistent application of prednisone [5,6].

Histological Elucidation

Upon microscopic examination, hyperplastic, keratinized, stratified squamous epithelium exhibits prominent hyper-parakeratosis, significant acanthosis and spongiosis [6,7]. Characteristically, epithelial cells display clear cytoplasm along with perinuclear eosinophilic condensation. Prominent hyperkeratosis, parakeratosis and significant acanthosis with cytoplasmic clearing of spinous epithelial cells are commonly discerned features. Distinctive, eosinophilic, intracellular, perinuclear condensation is delineated within superficial epithelial layers [6,7]. Upon morphological examination, superficial parakeratosis, acanthosis, and spongiosis is observed. Besides, perinuclear, eosinophilic condensation appears incorporated within epithelial cells. Minimal infiltration of mature lymphocytes is exemplified, which are confined to the stroma [6,7]. Characteristic histological features of white sponge nevus are enunciated as acanthosis, hyperkeratosis, parakeratosis and vacuolization of keratinocytes. Typically, perinuclear condensation of eosinophilic substance is observed [6,7]. Upon ultrastructural examination, the perinuclear eosinophilic substance is composed of tangled aggregates of keratin tono-filaments [6,7] (Figure 1-8).

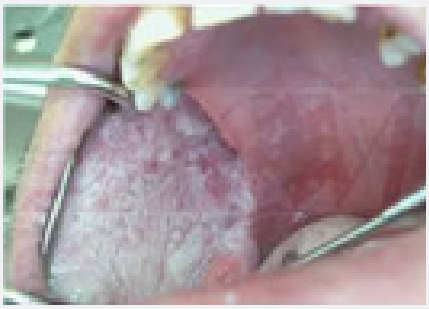

Figure 1: White sponge nevus exhibiting diffuse, thick, whitish, corrugated plaques incriminating the buccal mucosa [9].

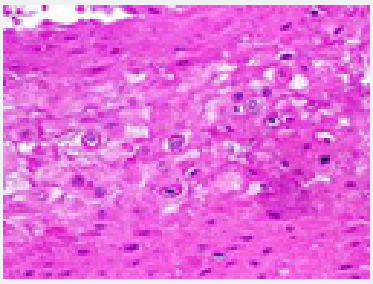

Figure 2: White sponge nevus exemplifying hyperkeratosis, parakeratosis, mild spongiosis, acanthosis and a perinuclear, eosinophilic condensation of fibrils [10].

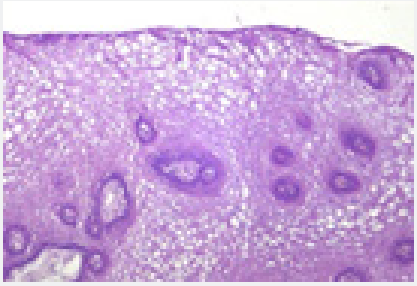

Figure 3: White sponge nevus enunciating acanthosis, hyperkeratosis, parakeratosis and eosinophilic, perinuclear condensation of fibrils with clearing of cytoplasm [11].

How to cite this article: Anubha B. The Pallid Plat-White Sponge Nevus. Theranostics Brain, Spine & Neural Disord 2022; 4(3): 555636. DOI: 10.19080/JOJS.2022.04.555636

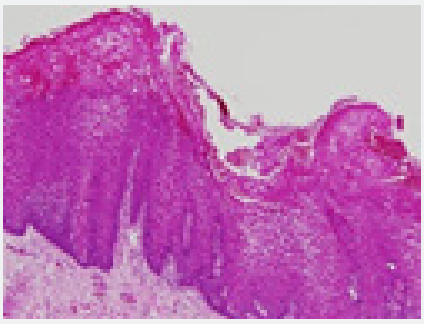

Figure 4: White sponge nevus delineating significant acanthosis, hyperkeratosis, parakeratosis and perinuclear aggregation of eosinophilic fibrils [12].

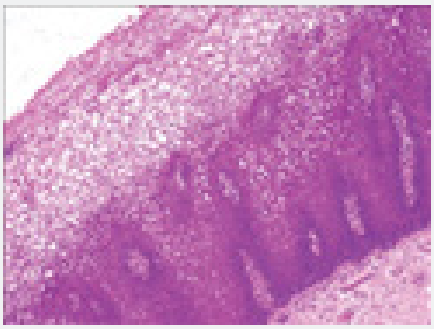

Figure 5: White sponge nevus depicting acanthosis, parakeratosis, spongiosis, cytoplasmic clearing and perinuclear accumulation of eosinophilic clearing [13].

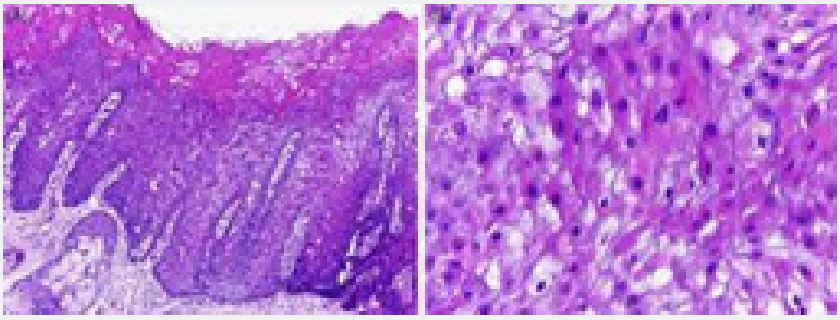

Figure 6: White sponge nevus displaying acanthosis, hyperkeratosis, parakeratosis and cytoplasmic clearing along with condensation of perinuclear fibrils [14].

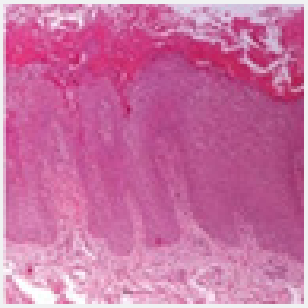

Figure 7: White sponge nevus demonstrating marked acanthosis, hyperkeratosis, parakeratosis and intracytoplasmic accumulation of keratin filaments [15].

How to cite this article: Anubha B. The Pallid Plat-White Sponge Nevus. Theranostics Brain, Spine & Neural Disord 2022; 4(3): 555636. DOI: 10.19080/JOJS.2022.04.555636

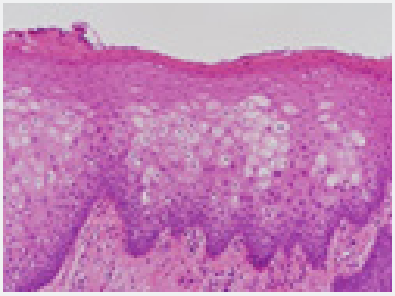

Figure 8: White sponge nevus delineating marked acanthosis, parakeratosis, hyperkeratosis and cytoplasmic clearing along with condensation of perinuclear tonofilaments [16].

Differential Diagnosis

White sponge nevus simulates oral lesions manifesting diffuse white plaques. Oral mucosal white lesions appearing in childhood or congenital disorders such as leukoedema, follicular keratosis, dyskeratosis congenita, hereditary benign intraepithelial dyskeratosis and oral lesions of pachyonychia congenita or Darier's disease require a segregation [3,4].

Generally, demarcation is required from conditions such as

a) Hereditary benign intraepithelial dyskeratosis which is associated with an absence of bilateral limbal conjunctival plaques [3,4].

b) leukoplakia may delineate a diffuse margin which blends into the circumscribing mucosa. However, variants of verrucous leukoplakia or proliferative verrucous leukoplakia exemplify a sharply defined lesion perimeter. Usually, history of tobacco consumption is absent [3,4].

c) Plaque type lichen planus and lupus erythematosus typically enunciate minimal striae upon periphery of white plaques [3,4].

d) Pachyonychia congenita, Darier's disease and dyskeratosis congenita demonstrate characteristic nail and cutaneous lesions. Oral lesions of Darier's disease display a cobblestone appearance [3,4].

e) Hyperplastic candidiasis with infection by Candida albicans may be diagnosed by cytological smears stained by periodic acid Schiff's (PAS) stain. Symptomatic lesions of white sponge nevus may be superimposed with Candida infection. Thus, oral candidiasis can be excluded by pertinent examination for fungal organisms and optimal therapeutic response to antifungal agents [3,4]. Additionally, differentiation is necessitated from conditions as oral lichen planus, syphilitic glossitis, chronic cheek biting, tobacco pouch keratosis, verrucous carcinoma and squamous cell carcinoma [3,4]. Acquired conditions as focal epithelial hyperplasia and oral florid papillomatosis emerging secondary to infection with human papillomavirus mandate distinction [3,4]. Leukoedema and hereditary benign intraepithelial dyskeratosis may demonstrate morphological alterations identical to lesions of white sponge nevus [3,4].

Investigative Assay

History of contributory factors as tobacco consumption or chemical burns is usually absent [7,8]. Upon cytological examination or staining with periodic acid Schiff's (PAS) stain, fungal hyphae may not be detected [7,8]. Exfoliative cytological smears stained with Papanicolaou technique exhibit an eosinophilic, perinuclear condensation [7,8]. Therapeutic Options The benign, asymptomatic, harmless condition is devoid of potential for malignant transformation and usually does not necessitate therapeutic intervention [7,8]. Generally, asymptomatic instances do not require treatment. Although inefficacious, diverse therapeutic agents employed are vitamins as beta-carotene, localized application of retinoic acid, antihistamines, tetracycline mouth rinse or antibiotics such as penicillin or azithromycin. Additionally, surgical excision and laser ablation of the lesion may be adopted. Lesion reoccurrence is usually absent at a follow up of 2 years [7,8]. Contemporary instances of white sponge nevus with associated genetic mutations may be subjected to genetic diagnosis and gene therapy at an optimal period [7,8].

References

1. Hyde JN (1909) An unusual naevus of the tongue in a five-year-old boy J Cutan Dis 27: 256.
2. Cannon AB (1935) White sponge nevus of the mucosa (nevus spongiosus albus mucosa). Arch Dermatol Syphilol 31(3): 365–370.
3. Elfatoiki FZ, Capatas S, Hayat DS, Fouzia H, Hicham A, et al. (2020) Oral White Sponge Nevus: An Exceptional Differential Diagnosis in Childhood. Case Rep Dermatol Med 2020.
4. Regezi JA, Scuibba JJ, Richard CKJ (2012) Oral Pathology Clinical Pathologic Correlations. 6th. Philadelphia, PA, USA: Saunders Elsevier, pp. 80-81.
5. Liu X, Li Q, Gao Y, Song S, Hua H (2011) Mutational analysis in familial and sporadic patients with white sponge nevus. Br J Dermatol 165(2): 448-451.
6. Sobhan M, Alirezaei P, Farshchian M, Gholamreza E, Khezrian L, et al. (2017) White sponge nevus: report of a case and review of the literature. Acta Med Iran 55(8): 533-535.

How to cite this article: Anubha B. The Pallid Plat-White Sponge Nevus. Theranostics Brain, Spine & Neural Disord 2022; 4(3): 555636. DOI: 10.19080/JOJS.2022.04.555636

7. Sanjeeta N, Nandini D, Takhellambam P, Sumita B (2016) White sponge nevus: report of three cases in a single family. J Oral Maxillofac Pathol 20(2): 300-303.

8. Quintella C, Janson G, Luciana RA, Damante JH (2004) Orthodontic therapy in a patient with white sponge nevus. Am J Orthod Dentofacial Orthop 125(4): 497-499.

DOI: 10.19080/JOJS.2022.04.555636

How to cite this article: Anubha B. The Pallid Plat-White Sponge Nevus. Theranostics Brain, Spine & Neural Disord 2022; 4(3): 555636. DOI: 10.19080/JOJS.2022.04.555636