# Nevus Sebaceus of Jadassohn on the Face with Full Thickness Surgical Excision: A Case Report

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Abstract: <u>Background</u>: Nevus sebaceus of Jadasshon (NSJ) is a quite rare inherited disorder that can cause cosmeutical problem. This case is the second case in the last 2 years at our center. Lesion of NSJ most frequently on the scalp (59.3%) but has also been found on the face (32.6%). Full thickness surgical excision is the choice of treatment. <u>Case</u>: A case of nevus sebaceus of Jadasshon, in 24 years old woman, with a brownish lump that is painless ang does not itch on the left cheek that has been present since birth. History of bleeds was denied. There are no abnormalities of the central nervous, skeletal or ocular systems. There is no family history of the same disorder. Dermoscopic feature: grayish yellow globules papillary appearance in "cobblestone pattern". Histopathological examination shows stratified squamous epithelium with hyperplasia, papillomatosis, and acanthosis. The dermis layer contains sebaceous glands, sudorifera glands, hair follicles, lymphocyte cell, periadnexal plasma cells. Patient was treated with full thickness surgical excision. <u>Discussion</u>: Lesions of NSJ are usually appears on the scalp and the shape of the lesion depends on the stage. In this case, the lesion accordance to the adult stage. There is hyperplasia of the lesion that leads to a larger and more verrucous appearance. Surgical excision was performed in this patient, although signs of malignancy and systemic involvement were not found. Based on the literature, although the percentage is low, there is a possibility that it will turn into malignancy in the future.

Keywords: congenital nevus, yellowish appearance, sebaceous glands

#### 1. Introduction

Nevus sebaceus of Jadassohn is a congenital abnormality first described by the dermatologist Josef Jadassohn in 1895. Nevus sebaceous of Jadasshon (NSJ) is a rare congenital skin lesion with unknown etiology that occurs in less than 1% of neonates. Nevus Sebaceus of Jadassohn is a benign hamartoma of the skin, characterized by hyperplasia of the epidermis, immature hair follicles and sebaceous and apocrine glands. Although NS can manifest on any part of the body, studies estimate 95% to 99% occur in the head and neck region.<sup>1-3</sup>

During infancy, NSJ typically appears as a smooth or velvety yellow-orange wellcircumscribed plaque. At puberty, hormonal changes cause proliferation and hyperplasia of the lesion and lead to a larger and more verrucous appearance that can become quite large. Later in life, the lesions can develop benign or malignant appendageal tumors that cause further disfigurement.<sup>4</sup>

In infancy or early childhood, nevus sebaceus histologically features immature and abnormally formed pilosebaceous units. At this stage of development, epidermal changes may reveal some acanthosis and mild papillomatosis. During puberty, the lesions enlarge clinically as well as microscopically. The sebaceous glands become much more prominent and are located unusually high in the dermis. The number of sebaceous lobules and malformed ducts increases. The hair follicles are typically immature vellus hairs rather than terminal hairs. At this growth phase, the epidermis becomes more papillated and acanthotic.<sup>5</sup>

The timing of excision is also a matter of debate; with some advocating early excision and others suggesting that delayed excision is reasonable. The definitive treatment of NS consists of full-thickness operative excision. Given the tendency of NS to expand during puberty, some argue that excision should be performed before this time to decrease the extent of excisional surgery and to improve the overall cosmetic result. Although current recommendations for surgical excision include removal of an NS on a cosmetically sensitive area or to avoid future unsightly growth of the lesion there is no agreed upon protocol describing surgical approach or timing of excision.<sup>6</sup>

#### 2. Case Report

A case of 24-years-old woman who complained brownish lump on the left cheek with no itchy and pain that have been increasing since birth. Initially the lump is not thick and yellowish-brown in color. But after puberty, the lump gets thicker with a bumpy surface and becomes more brown in color. There were no history of trauma around the lesion, spontaneous bleeding, seizures and headache. There were no history of facial and skeletal muscle weakness, hearing loss, reduced vision and cataract.

On physical examination, generalized state was in normal limit. From dermatology state, there was lession at left lateral cheek, localized, size 3,5 cm x 1 cm x 1 mm, efflorescence skin color-brownish verucous plaque. The

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**Figure (a):** before excision, skin color-brownish plaque verucous on the left lateral cheek 3,5cm x 1 cm x 1mm. Figure (b) (dermoscopic features) : grayish yellow globules papillary appearance in "cobblestone pattern", teleangiectasis (-). Figure (c) (histopathological examination). Sebaceus glands opening directly into epidermis, achantosis, papilomatosis. Figure (d): Post surgical excision after 45 days

result from the laboratory finding was in normal limit. Dermoscopy features we found grayish yellow globules papillary appearance in "cobblestone pattern" and teleangiectasis was negative. From histopathological examination, there were pieces of tissue with a surface covered with stratified squamous epithelium with hyperplasia, papillomatosis, and acanthosis. The dermis layer contains sebaceous glands, sudorifera glands and hair follicles. Visible as a lymphocyte cell, periadnexal plasma cells.

Based on the all result examination we made diagnosis with nevus sebaceous of Jadasshon (NSJ) and patient we plan to do full thickness surgical excision.

## 3. Discussion

Nevus sebaceus of Jadassohn (NSJ) is a hamartomatous lesion with an epithelial and adnexal origin which is typically asymptomatic at birth. Sebaceus nevus is a benign lesion that occurs most frequently on the scalp (59.3%) but has also been found on the face (32.6%), preauricular area (3.8%), neck (3.2%), and locations off the head and neck (1.3%). Nevus sebaceus of Jadassohn (NSJ) evolves through three stages. In the infantile stage, it presents as solitary smooth, yellow, well-circumscribed hairless plaques. At puberty, it becomes more prominent as a verrucous or mamillated appearance. The final stage is characterized by or nodular tumoral appearance with peripheral telangiectasias.3,7

In this case from dermoscopic examination, we found yellowish globules-grayish papillary appearance in "cabblestone pattern". This is in accordance with the literature from Kelati (2017) about dermoscopic analysis study 13 cases of NSJ. In this study, in addition to the yellow-grayish papillary appearance, there were also features found yellowish globules aggregated in a "cobblestone pattern," whitish-yellow lobular aspect, homogenous yellowish, and peripheral vascularization (linear, irregular, or arborescent).<sup>8</sup> In this case, we report dermoscopic features of an isolated lesion of nevus sebaceus on face without involvement of scalp, which is quite rare. As there is a risk of developing secondary benign or malignant neoplastic changes in NSJ, so careful monitoring is required

for its early detection and dermoscopy is a useful tool for this monitoring without invasive procedure like biopsy.

The differential diagnosis of nevus sebaceus depends on the stage of development. In early infancy, the differential diagnosis may include aplasia cutis congenita or mastocytoma. Other differential diagnoses in later stages of development include epidermal nevus, seborrheic keratosis and verruca.<sup>9</sup> In this case, from histopathology examination, this case revealed verrucous proliferation of epidermis and dermis showed ill-developed hair follicles, mature sebaceous glands with inflammation cell. On the basis of clinical, histopathological, and dermoscopic findings, a diagnosis of nevus sebaceous was made and patient will plan to removal of the lesion with surgical excision full thickness.

Nevus sebaceus is a clinical diagnosis; additional workup is typically unnecessary. The exception would be a child with nevus sebaceus syndrome that presents with a large or multifocal lesion that may be associated with extracutaneous involvement of the ocular, skeletal, or central nervous systems. In this case, referral to neurology and other specialists may be indicated if other physical anomalies are present.<sup>9</sup>

Individuals with NS may develop benign or malignant tumors, with increasing evidence supporting the former to be the majority. A meta-analysis of 4900 cases found secondary tumor development in 24% of patients, most commonly benign basaloid proliferations such as trichoblastomas. Malignant transformations have been reported with an average incidence of 8% and ranging from 0% to 22%, most commonly basal cell carcinoma, although there is some controversy of falsely high estimations, given the misdiagnosis of trichoblastomas that appear histologically similar to basal cell carcinoma in earlier reports. Their incidence increases with the age, and malignant growths have not been observed before the age of 10 years.<sup>6,10</sup> In our case the Patient has 24 years old and there was no history of any other cutaneous and systemic involvement.

The definitive treatment of nevus sebaceous is full-thickness excision. However, the necessity and timing of excision to prevent possible future malignancy are still debated. The decision to excise the lesion should be made in individual patients, based upon age, extension and location of the

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lesion, and the patient's or parents' concern about the cosmetic appearance and/or risk of malignancy. Since the risk of malignant transformation appears to be lower than previously believed, observation may be reasonable for lesions that do not cause cosmetic concern.<sup>10</sup> In this case, although there were no signs of malignancy, the patient was treated with surgical excision. Therapeutic management of this case includes full-thickness excision with clear margins.

Alternatives to surgical excision include photodynamic therapy, carbon dioxide laser resurfacing, and dermabrasion. However, since these treatment modalities do not completely remove the lesion, the risk of recurrence and potential for neoplasm development remains.<sup>6</sup>

## 4. Conclusion

We report a case of Jadasshon nevus on the face in a 24year-old woman. These cases are quite rare and are usually the site of the lesion on the scalp. The lesion in this case was an adult stage lesion with efflorescence of brown verucous plaque and a dermoscopy yellowish-brown papillary appearance. Full thickness surgical excision treatment for this patient although signs of malignancy and systemic involvement were not found because according to the literature, although the percentage is low, there is a possibility that it will turn into malignancy in the future.

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