



## Nevus Sebaceous Of Jadassohn: Diagnosis & Management of A Rare Case

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### Abstract

Nevus sebaceous is a rare hamartomatous lesion usually diagnosed on histopathology. There is a marked proliferation of sebaceous glands along with other adnexal units. We report a case of Nevus sebaceous in an 18-year-old male who presented with a pigmented lesion on left ala of nose. Initially the patient was sent for fine needle aspiration cytology (FNAC) and hence a possibility of Nevus sebaceous of Jadassohn was given. However, to confirm the diagnosis, a detailed histopathological evaluation was done from the excised specimen. Nevus sebaceous has been documented with presence of underlying benign or malignant lesion, where in lies the importance of histopathology to rule out such lesion, if any. However, in our case no underlying lesion was found. The patient was followed up for a year and no recurrence was noted.

**Keywords:** Sebaceous hyperplasia, histopathology, FNAC, melanin.

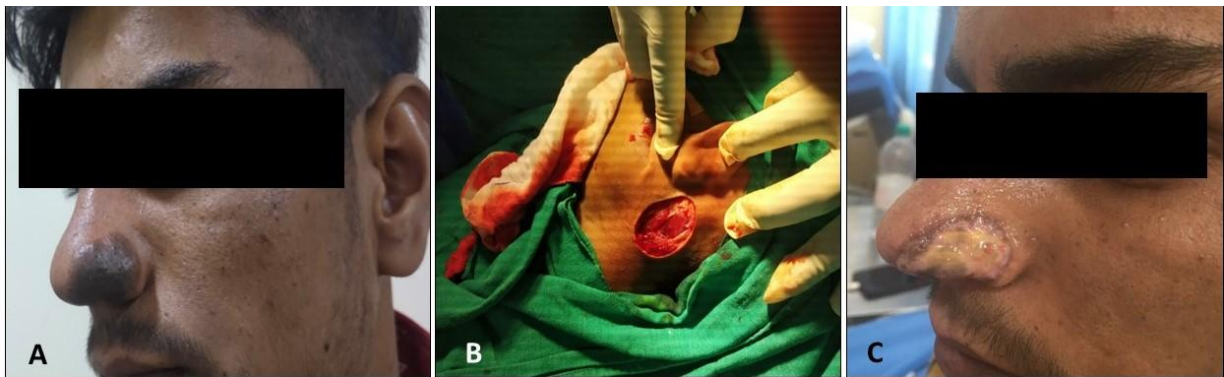
### Introduction

Nevus sebaceous of Jadassohn (NSJ) is an uncommon hamartoma comprising of sebaceous glands along with proliferation of sweat glands and hair follicles.<sup>1-2</sup> Jadassohn was the first person to describe this entity in 1895 calling it as a circumscribed hamartoma predominantly consisting of sebaceous units.<sup>1-3</sup> Morphology of this lesion varies as per the duration of the lesion.<sup>3-5</sup> Few of secondary tumors have been found to be in association with nevus sebaceous.<sup>2-5</sup> Hence, an extensive histopathological evaluation is required to rule out other differential diagnosis and the presence of any benign or malignant lesion in association with nevus sebaceous. We describe a case of 18-year-old

male with blackish lesion on left ala of nose for the last 10 years and was diagnosed as NSJ based on cytological examination, followed by histological evaluation which ruled out any associated underlying pathology.

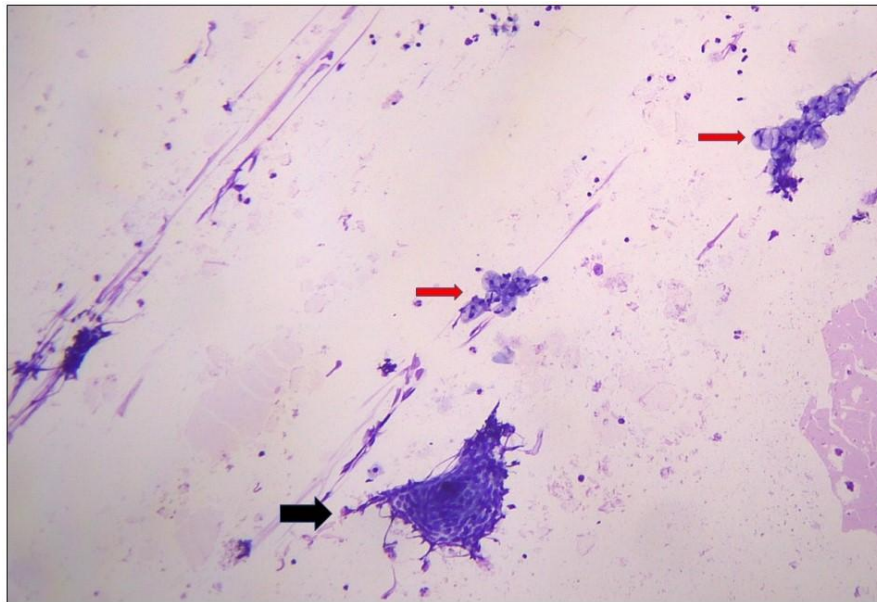
### Case Report:

An 18-year-old male presented with a pigmented swelling over the left ala of nose for the last 10 years [Figure 1A]. The patient had a history of trauma at the same site 10 years back. On examination, a brownish- black swelling was noted on the left ala of nose, measuring 3x4 cms. Margin of the swelling was slightly elevated from the adjacent tissue. No central depression was noted.

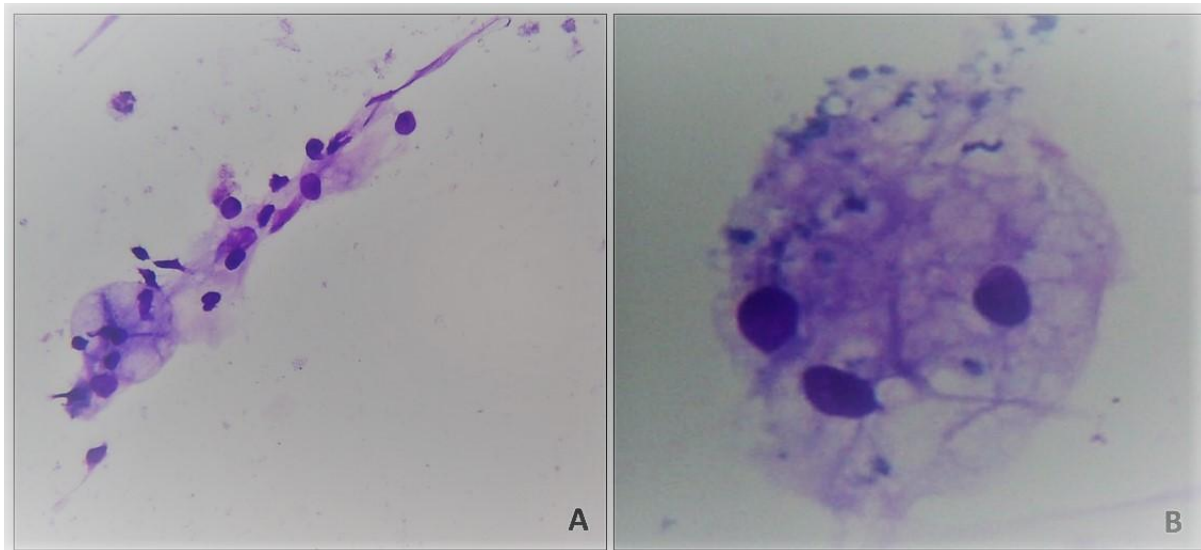


**Figure 1A-C: A: Image showing a pigmented lesion over left ala of nose; B: Full-thickness skin graft taken from left post-auricular region to correct the defect; C: Well-taken graft on follow-up.**

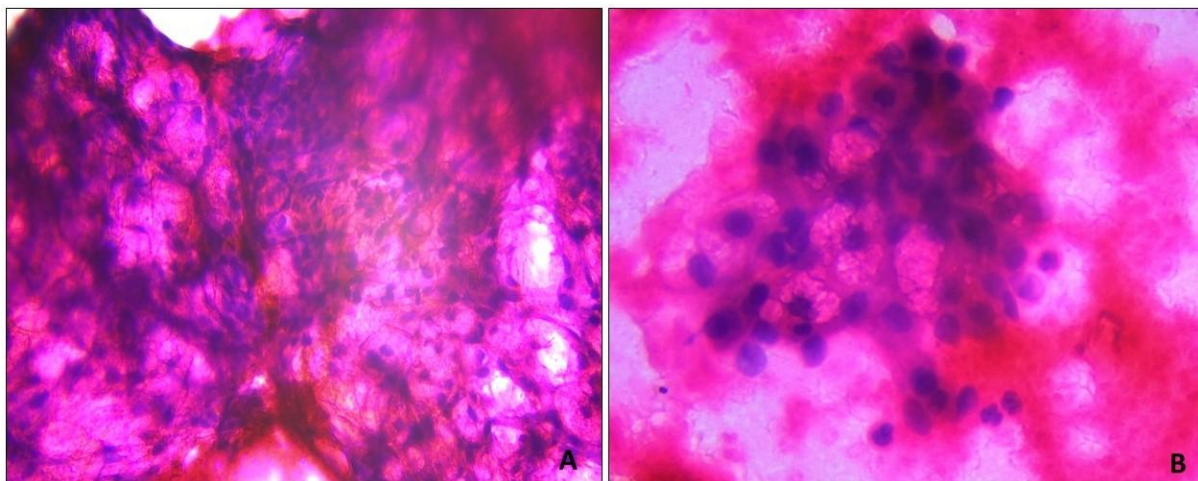
The patient was sent for FNAC. Blood mixed aspirate was obtained, and smears examined show sheets & clusters of sebaceous and basaloid cells [Figure 2-4]. At places, both the components were seen intermixed. Based on cytological evaluation, a possibility of Nevus sebaceous of Jadassohn was given. However, to confirm the diagnosis and rule out sebaceous hyperplasia and other differentials a thorough histopathological examination was advised, which would also help in ruling out any underlying benign or malignant lesions.



**Figure 2: Low power view of FNAC smear showing clusters of sebaceous cells (red arrows) and sheet of basaloid cells (black arrow) [Pap, 20X]**



**Figure 3A-B: Smears showing groups of sebaceous cells with vacuolated cytoplasm, bland nuclei and fine granules (? Pigment) [Pap, A-20X, B-40X]**



**Figure 4A-B: H&E-stained cytology smears showing the finely vacuolated sebaceous cells and basaloid cells in clusters [H&E, A& B- 40X]**

The lesion over left ala of nose was excised and sent for histopathological evaluation [Figure 5A]. The defect in the nose was corrected by taking full-thickness skin graft from the left post-auricular region [Figure 1B]. The graft was well taken and vascularized in subsequent days of follow-up [Figure 1C].

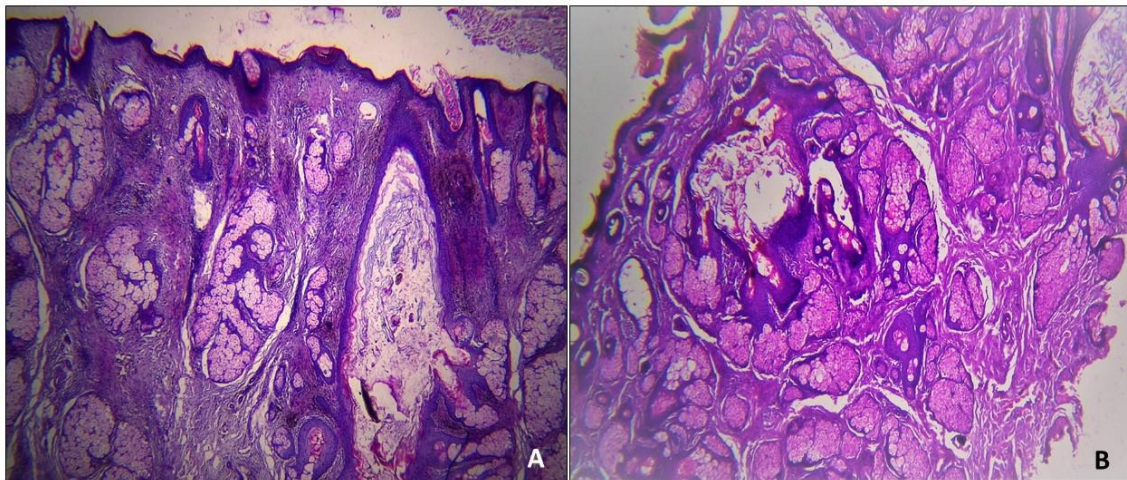
On gross, a flattened skin covered tissue measuring 4x3.5 cms was received. External surface showed blackish discoloration measuring 3.5x2.5 cms [Figure 5B]. Focal areas showed presence of hair. Under surface of the skin revealed grey-white and focal grey-brown areas [Figure 5C].



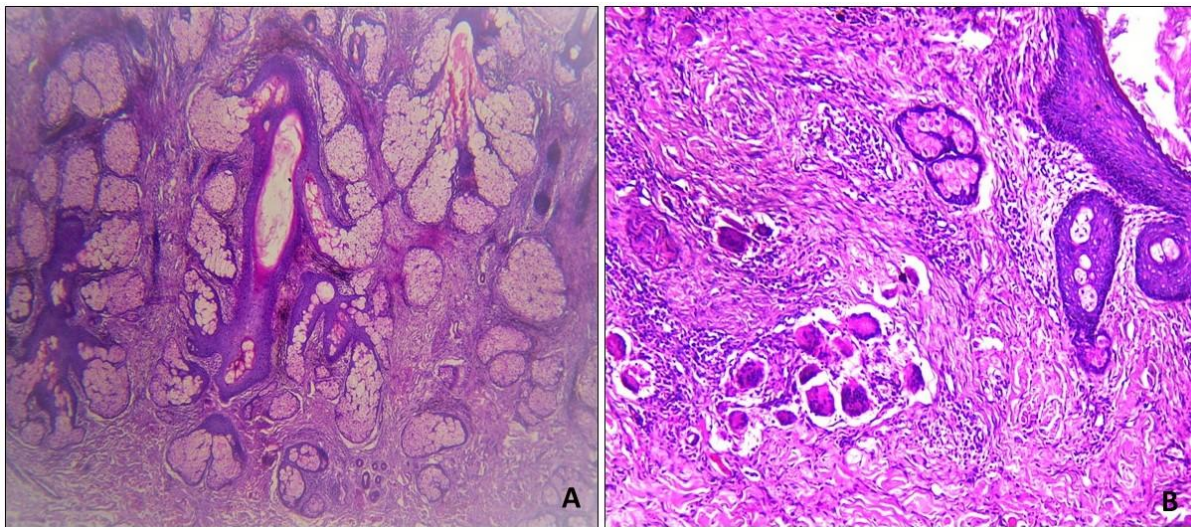
**Figure 5A-C: A- Surgically excised specimen, B-C: Gross image showing the 4x3.5 cms, pigmented flattened skin & hair covered, flattened tissue mass.**

Sections examined showed hyperkeratosis and mild papillomatosis [Figure 6A-B]. Sub-epidermal zone showed abnormal hair follicles along with pilosebaceous units. Extensive sebaceous hyperplasia was noted [Figure 7A]. At places melanin pigment was seen in stroma along with small collection of

eccrine glands & giant cell reaction [Figure 7B]. Histopathological features were consistent with nevus sebaceous. However, in our case no underlying lesion was found. The patient was followed up for a year and no recurrence was noted in the patient.



**Figure 6 A-B: Sections showing hyperkeratotic epidermis with mild papillomatosis and showing marked proliferation of sebaceous units [H&E, A & B-20X]**



**Figure 7A-B: A- Sections showing sebaceous hyperplasia along with dilated duct [H&E, 40X]; B- Focal area showing giant cell reaction with eccrine glands [H&E, 20X]**

### Discussion:

Nevus sebaceous of Jadassohn (NSJ) is a benign lesion which is usually asymptomatic at birth.<sup>1-3</sup> It has an epithelial and adnexal origin. This hamartomatous lesion is usually seen on the scalp, followed by face, pre-auricular area, neck region.<sup>2,4</sup> The lesion has a varied morphologic appearance as per the age of lesion. It evolves through 3 different stages.<sup>3-6</sup> The first is the infantile stage which presents as a solitary, smooth, yellow well-circumscribed hairless plaque. It becomes more prominent as a verrucous like in the second stage which is usually seen around puberty. The third and the final stage is characterized by nodular or tumoral appearance with peripheral telangiectasia.

This lesion is associated with few secondary benign and malignant tumors. Usually, it is noted that the association of these secondary tumors increase after puberty.<sup>3-7</sup> Underlying secondary tumors could be trichoblastoma, syringocystadenoma papilliferum, trichilemmoma, sebaceoma, nodular hidradenoma, hidrocystoma and eccrine poroma. Very rarely, malignant cutaneous neoplasm can be seen like basal cell carcinoma along with squamous cell carcinoma, trichilemmal carcinoma, sebaceous carcinoma, porocarcinoma and apocrine carcinoma.<sup>4-9</sup> Histopathological examination becomes very important in diagnosis of NSJ and the underlying secondary tumors if any.

Histopathology presents a wide morphologic appearance in all the three clinical stages.<sup>6-9</sup> There is

a slight acanthotic and pigmented epidermis in the infantile, i.e., patch/ plaque stage. The hair follicles are small, not formed completely and appears as solid cords of undifferentiated basaloid cells. However, in this stage, the sebaceous glands are prominent. In a study of 20 cases of NSJ, author noted the absence of normal terminal hair follicles within the lesion although it can be seen in the rest of epidermis.<sup>5</sup>

The second stage, i.e., elevated or verrucous, which is seen around the adolescence, shows an increase in the thickness of the lesion demonstrated by smooth surface nodularity or verrucous hyperkeratosis. Histology shows verrucous epidermal hyperplasia which can also be seen in epidermal nevus. However, it can be differentiated from epidermal nevus by the presence of malformation of dermis showing prominent hyperplasia and malpositioning of sebaceous glands. Hair follicles are small and primordial. There is marked increase in sebaceous glands.

The nodular/ tumoral stage (final stage) is seen in adulthood and shows epidermal hyperplasia, large sebaceous glands and ectopic apocrine glands. This stage has to be differentiated from other benign and malignant adnexal tumor.

The various histopathological features noted in NSJ in a study by Neerja Puri<sup>5</sup>, were immature hair follicle in 20 cases (100%), followed by acanthosis in 16 cases (80%), papillomatosis in 16 cases (80%), dilated keratin filled infundibulum in 5 cases (25%), immature sebaceous glands in 4 cases (@0%),

eccrine gland hyperplasia in 3 cases (15%). Overall, we can see that, in patch/ plaque stage, there is immature sebaceous glands and hair follicles localized in deeper dermis. In the elevated/ verrucous plaque stage, there is immature and mature sebaceous gland with inflammatory infiltrate. This is seen localized between reticular and papillary dermis. Mature sebaceous glands are seen in the superficial dermis in the nodular/ tumoral stage. Sahu et al<sup>9</sup> described a detailed histological & dermoscopic features in a patient of NSG on face.

A very common entity which should be differentiated from NSJ is nevoid sebaceous hyperplasia.<sup>5,6-9</sup> Clinically, nevoid hyperplasia presents as multiple skin coloured to yellowish papules with a central depression with face being the common location. There is no change in the surface of the lesion. Histologically, nevoid hyperplasia, shows an increase in proliferation of sebaceous glands near to epidermis and is composed of mature sebaceous units with ducts opening directly onto the skin surface.

On the other hand, NSJ is usually seen as a flat, smooth surfaced, solitary plaque-like lesion, mostly noted on the scalp region. Gradually, the surface becomes irregular and verrucous at adolescence. NSJ shows mild epidermal hyperplasia, with abrupt absence of mature hair follicles, immature sebaceous units and few ectopic apocrine ducts. In the verrucous stage around adolescence, many irregularly arranged sebaceous glands are seen superficially, with ectatic ducts opening into the epidermis.

Risk of developing secondary benign/malignant lesion is association with NSJ makes it important to carefully monitor the patient with a detailed histopathological evaluation. The treatment includes surgical excision, CO2 laser, photodynamic therapy, cryotherapy and electrosurgery.<sup>7-9</sup>

### Conclusion:

Nevus sebaceous of Jadassohn (NSJ) is a hamartomatous lesion comprising of sebaceous units

along with other adnexal structures. There is absence of normal terminal hair follicles in the lesion. We present a rare case of NSJ in young male, which was initially diagnosed on FNAC adding to the rarity of our case. NSJ is usually associated with an underlying secondary tumor, hence a detailed histopathological evaluation of excised specimen is of utmost importance to rule any benign or malignant lesion, if any.

### References:

1. Sato T, Tanaka M. Linear sebaceous hyperplasia on the chest. *Dermatol Pract Concept* 2014;4:93-5.
2. Nair PA, Diwan NG. Sebaceous hyperplasia mimicking linear wart over ear. *Int J Trichology* 2015;7:170-2.
3. Mandal RK, Das A, Chakrabarti I, Agarwal P. Nevoid sebaceous hyperplasia mistaken as nevus sebaceous: Report of four cases. *Indian J Dermatol Venereol Leprol* 2017;83:213-6.
4. Cribier B, Scrivener YG. Tumours arising in nevus sebaceous: A study of 596 cases. *J Am Acad Dermatol* 2000;42:263-8.
5. Puri N. A clinical and histopathological study of nevus sebaceous. *J Pakistan Association of Dermatologists* 2014;24(1):31-3.
6. Alsaad KO. Skin adnexal neoplasms-Part-I: An approach to tumours of pilosebaceous unit. *J Clin Pathol* 2007;60:129-44.
7. Jonathan SD. Epidermal nevus. *Dermatol Online J* 2001;7:14.
8. Verma KK, Ovung EM. Epidermal and sebaceous nevi treated with carbon dioxide laser. *Indian J Dermatol* 2002;68:23-4.
9. Sahu P, Lakra S, Dayal S. Nevus sebaceous on face: Histopathological and dermoscopic correlation. *Indian Dermatol Online J* 2020;11:878-80.