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Case Report

Ear, Nose and Throat Section

Nevus Sebaceous of Jadassohn Mimicking a Pilonidal Sinus of Forehead: A Rare Case Presentation

PARINDITA SARMAH[1], PRASAD DESHMUKH[2], SAGAR GAURKAR[3]

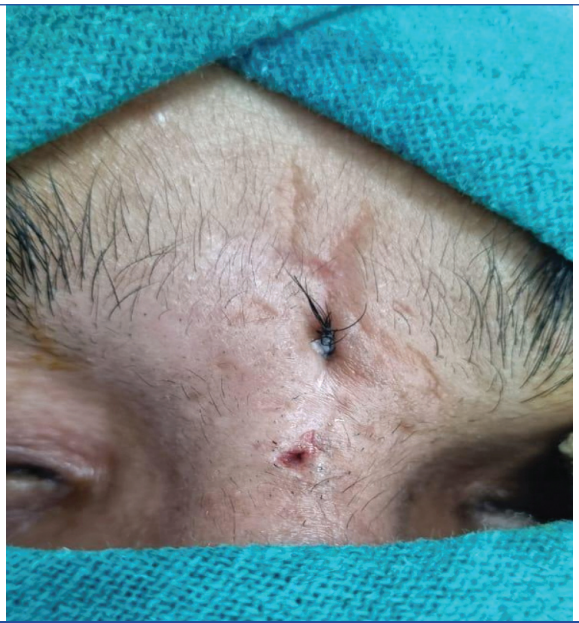
ABSTRACT

Nevus Sebaceous of Jadassohan (NSJ) is a hamartoma that presents as a cutaneous lesion, which can be considered an overgrowth of sebaceous glands, hair follicles, and other epidermal structures. Although it is usually a benign condition, the lesion is capable of producing secondary neoplasms, both benign and infrequently, malignant. This case report discusses a 21-year-old male patient who presented with a lesion on the forehead, manifesting as a pilonidal sinus due to the presence of a tuft of hair at the end of the sinus tract following childhood trauma. The presence of a sinus tract with hair growth in the glabellar region is a rare and diagnostically challenging presentation of NSJ. This case highlights the complexities of managing such chronic wounds and the potential complications arising from trauma.

Keywords: Computed tomography sinogram, Glabella, Sinus tract, Trichoblastoma

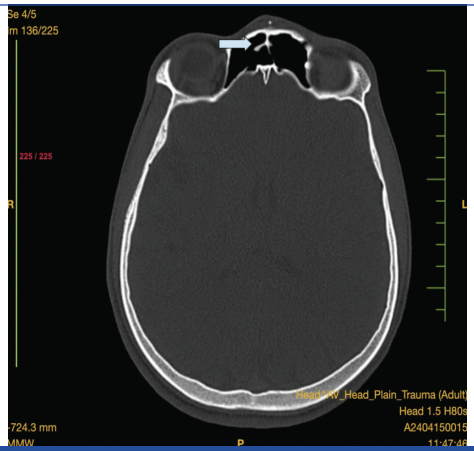
CASE REPORT

A 21-year-old male presented to the Department of Otorhinolaryngology with a lesion on his forehead, initially resembling a pilonidal sinus due to the presence of a tuft of hair at the end of the sinus tract, which had been present since the age of two. He revealed a history of traumatic laceration to the glabellar region at that age, which was followed by the application of sutures in that area. The wound did not heal properly, resulting in pus discharge for six months. During that period, another episode of trauma to the same area occurred, causing persistent purulent discharge from the region for almost three months [Table/Fig-1]. The patient was treated with intravenous antibiotics, and timely dressing of the wound site was performed to promote faster healing. After three months, active discharge from the wound had ceased, and only a tuft of hair has been emerging from the glabellar region since then. There is currently no association with pain, swelling, redness, or an increase in temperature in that area.



[Table/Fig-1]: A discharging sinus with a tuft of hair present at the forehead.

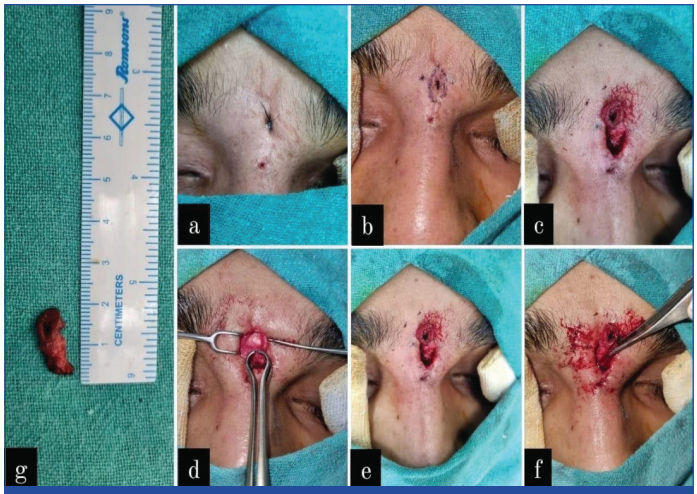
The patient underwent a Contrast Enhanced Computed Tomography (CECT) sinogram, which showed a linear tract extending from the skin into the subcutaneous tissue, reaching up to the upper end of the nasal bone. The scan indicated a maximum thickness of 1.7 mm over a length of 1.4 cm, suggestive of a sinus tract [Table/Fig-2].



[Table/Fig-2]: CT sinogram depicted a linear tract with a maximum thickness of 1.7 mm over a length of 1.4 cm that was suggestive of a sinus tract.

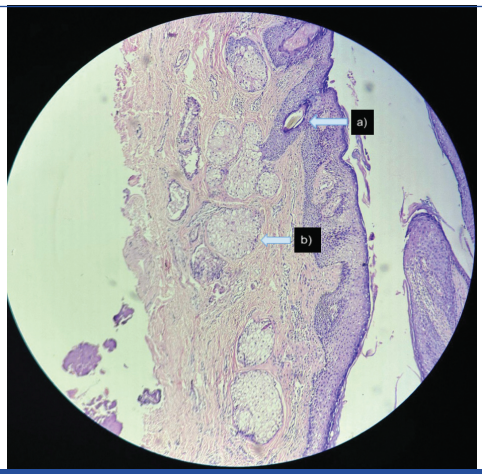
For a case of NSJ, particularly when presenting as a sinus-like lesion on the forehead, the differential diagnosis should include conditions that share clinical features such as the presence of a sinus tract, tuft of hair, or skin nodules. These conditions include pilonidal sinus, epidermal inclusion cyst, trichilemmal cyst (pilar cyst), dermoid cyst, and seborrhoeic keratosis. NSJ can occasionally undergo secondary changes, including benign adnexal tumours like Syringocystadenoma Papilliferum or, more rarely, malignant transformation into Basal Cell Carcinoma (BCC), underscoring the importance of long-term surveillance for patients diagnosed with this condition.

The patient was scheduled for sinus tract excision under local anaesthesia [Table/Fig-3]. The sinus tract on the forehead, near the nasal root, was first marked with methylene blue dye to enhance visualisation. An elliptical incision was then made around the opening of the sinus tract to allow for better access. Blunt dissection was carefully performed down to the nasal bone and up to a point above the rhinion, fully exposing the tract. The sinus tract, measuring approximately 2×0.5×0.2 cm, was completely excised and sent for Histopathological Examination (HPE) to assess any underlying pathology. The wound was closed with Prolene 3-0 suture material, and dressing with L/A T-bact was applied over the sutures.



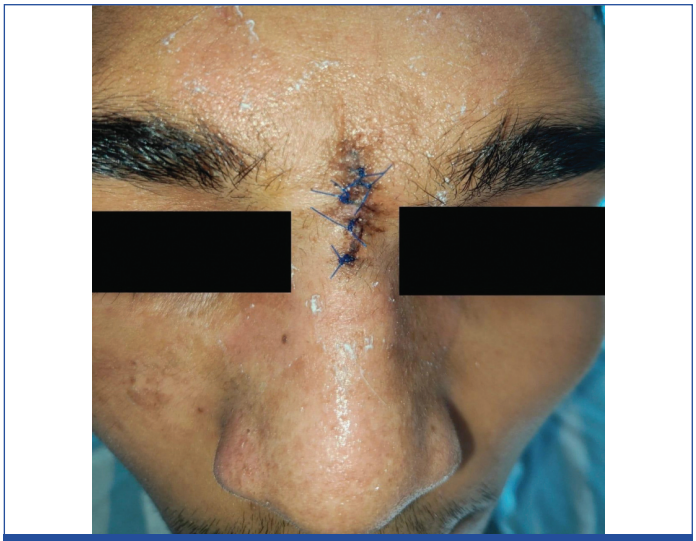
[Table/Fig-3]: a) Inspecting the sinus tract on the forehead; b) Marking done with methylene blue dye over the sinus opening at root of nose; c-e) Elliptical incision given around sinus tract opening and blunt dissection was carried out till the nasal bone and followed till above the level of rhinion; f) Sinus tract dissected completely and removed and sent for HPE; Sinus tract measuring around 2×0.5×0.2 cm in size.

The histopathology report described the sample as a single, greyish-white tissue piece with skin and hair attached, measuring 2×0.5×0.2 cm, suggestive of NSJ [Table/Fig-4]. Histopathological features of NSJ include epidermal hyperplasia, where the epidermis thickens and shows acanthosis, or an increased number of cells in the stratum spinosum. Sebaceous gland hyperplasia is a hallmark feature, with sebaceous glands appearing enlarged and more numerous, sometimes extending deeper into the dermis. Hair follicles may be underdeveloped or absent in the affected area. Additionally, apocrine glands may show hyperplasia or cystic changes. The stroma, or connective tissue, is often fibrotic and thickened, particularly in older lesions. These characteristic features distinguish NSJ from other conditions such as pilonidal sinus.



[Table/Fig-4]: Histopathology report of Nevus Sebaceous of Jadassohan (NSJ) revealing epidermal thickening, sebaceous gland hyperplasia, apocrine changes, and potential secondary changes within the lesion {Haematoxylin and Eosin (H&E), 10x}. a) Hair follicle; b) Hyperplasia of sebaceous glands.

Following the sinus tract excision, the patient was advised on standard postoperative care, including maintaining wound hygiene and keeping the site dry to promote healing [Table/Fig-5]. The surgical site was inspected regularly for any signs of infection, such as redness, swelling, or discharge. The patient was prescribed a seven-day course of a third-generation cephalosporin antibiotic, Tablet Cefixime 200 mg BD, along with analgesics and anti-inflammatory drugs like Tablet Zerodol SP BD and Tablet Chymoral Forte TDS to manage pain and prevent infection. Sutures were removed after 10 days. The excision site healed well during the follow-up visit, with no signs of recurrence or complications. The patient was counselled on the importance of periodic follow-ups to monitor the site for any future changes, particularly considering the potential for malignant transformation in such lesions.



[Table/Fig-5]: Postoperative picture of the lesion.

DISCUSSION

The NSJ, a unique skin lesion described by the dermatologist Josef Jadassohn in 1895 [1], is a congenital hamartoma composed of abnormal sebaceous glands, epidermal hyperplasia, and ectopic elements from adnexal structures. It typically appears on the scalp or face during infancy and can present as a waxy, hairless plaque [2]. Although NSJ is relatively uncommon, its occurrence on the forehead is even rarer, and its clinical presentation, which mimics a pilonidal sinus, adds an additional layer of diagnostic complexity [3].

Recent literature has expanded the understanding of NSJ's pathophysiology, with studies identifying genetic mutations, primarily in the Harvey Rat Sarcoma (HRAS) and Kirsten Rat Sarcoma (KRAS) genes, as major factors in lesion development. These mutations lead to abnormal cellular proliferation and signalling, which may increase the risk of secondary neoplasms over time, such as BCC, syringocystadenoma papilliferum, and rarely, sebaceous carcinoma [4]. However, trauma has also been suggested as a potential trigger for NSJ, as in this case, where trauma may have contributed to its onset [5].

A major challenge in diagnosing NSJ in this case was its unusual presentation, which mimicked a pilonidal sinus. Pilonidal sinus disease is typically found in areas prone to friction and hair entrapment, like the sacrococcygeal region, and rarely occurs on the forehead [6]. The lesion's sinus tract and the presence of hair initially led to a clinical diagnosis of pilonidal sinus, a reasonable assumption given its resemblance to this condition [7]. However, pilonidal sinus typically exhibits inflamed sinus tracts, granulation tissue, and foreign body reactions, which were not found in this case [8].

Histopathological examination was crucial, as it revealed features characteristic of NSJ, such as marked sebaceous gland hyperplasia and significant epidermal thickening [9]. Importantly, the examination ruled out a pilonidal sinus by the absence of features typically associated with it, such as granulation tissue, foreign body reactions, and inflamed sinus tracts. These findings collectively supported the diagnosis of NSJ and excluded the possibility of a pilonidal sinus, which would have required different management [10].

The uniqueness of this case lies not only in its unusual presentation but also in the location of the lesion on the forehead. A tuft of hair in Nasojugal Sinus (NSJ) is also atypical, as the lesion is usually hairless, further complicating the clinical diagnosis [11]. This case highlights the need for clinicians to consider NSJ as a differential diagnosis when encountering atypical sinus-like lesions on the face, particularly in cases where hair is present [12].

The management of NSJ generally involves surgical excision, particularly when lesions are large, symptomatic, or associated with

cosmetic concerns. In addition, complete excision is often advised due to the risk of secondary neoplastic changes [13]. In this case, excision of the sinus tract under local anaesthesia provided both therapeutic and diagnostic benefits, confirming the diagnosis while preventing further complications.

A review of similar cases in recent literature underscores the need for a broad differential diagnosis when evaluating such atypical lesions. Kumar P et al., demonstrated a similar case of NSJ over the posterior surface of the right ear, which had a whitish-grey plaque with multiple skin-coloured to dark brown soft whorled masses over it [14].

Silva MP et al., also presented a case of NSJ located at the posterior aspect of the left pinna since birth, which had a familial history of BCC. The lesion was surgically excised, and primary closure was performed [15].

Scott M et al., reported an interesting case of an 18-year-old Arab male who presented with a velvety plaque measuring 7 cm in length in his left preauricular area. Excision biopsy revealed NSJ with infundibular and keratinous cysts [16].

In this case report, the management of a nascent sinus junction (NSJ) on the forehead, which initially mimicked a pilonidal sinus, required a thorough clinical assessment, complete surgical excision, and histopathological confirmation for an accurate diagnosis. This unusual presentation underscores the need to consider NSJ in the differential diagnosis of sinus-like lesions. Postoperative follow-up is crucial to monitor for any recurrence or rare malignant transformation, ensuring patient safety and achieving favourable cosmetic outcomes over the long-term.

CONCLUSION(S)

In conclusion, this case highlights the need for a comprehensive diagnostic approach when evaluating sinus-like lesions in unusual locations, such as the forehead. Careful clinical assessment, complete surgical excision, and thorough histopathological examination were crucial in accurately diagnosing NSJ and ruling out other potential conditions, such as pilonidal sinus. The present case underscores the importance of including NSJ in the differential diagnosis of similar lesions to avoid misdiagnosis and ensure effective treatment. Postoperative follow-up remains essential, not only to monitor for recurrence but also to detect any rare malignant transformation, ultimately supporting patient safety and achieving a favourable cosmetic outcome.

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