Section: General Surgery

Case Report

CASE REPORT ON NASAL NEVUS SEBACEOUS: A SURGEON'S PERSPECTIVE

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ABSTRACT

Nevus sebaceous is a congenital, hamartomatous lesion characterized by the presence of sebaceous glands, hair follicles, and other skin appendages, typically located on the head or neck. It often presents as a yellowish, hairless, and velvety patch in early childhood and may undergo various changes with age, including the development of nodules or verrucous growths. This case report describes a rare presentation of nevus sebaceous in a pediatric patient, highlighting its clinical features, diagnostic evaluation, and management. The patient presented with an asymptomatic lesion on the nose that had gradually enlarged over time. Histopathological examination confirmed the diagnosis of nevus sebaceous, revealing a mixture of sebaceous glands, hair follicles, and epidermal hyperplasia. The report also discusses the potential for malignant transformation, which, although rare, can occur in long-standing cases. Treatment options, including surgical excision, are reviewed, with emphasis on the importance of monitoring for any changes suggestive of malignant conversion. This case underscores the need for awareness of nevus sebaceous in adult populations and the role of early intervention in preventing complications.

Keywords: Differentiation, Hamartoma, Nevus sebaceous, Jadassohn Disease II, Nose.

INTRODUCTION

Nevus sebaceous is a birthmark on the skin, typically affecting the sebaceous glands, sweat glands, hair follicles, and epidermis. A nevus sebaceous diagnosis is sometimes called "Jadassohn Disease II" or "organoid nevus." It commonly presents at birth or early childhood and most often occurs on the scalp but can appear on the scalp, forehead, face or neck. It has an incidence of 0.3% among the newborns. While usually benign, the lesion has a potential for secondary neoplastic transformation, particularly in adulthood.[1,2] The incidence of these tumours increases with age, particularly after puberty. They include benign adnexal tumours such as trichoblastoma, sebaceous, nodular hidradenoma, hidrocystoma, and eccrine poroma. Malignant cutaneous neoplasms are less commonly seen and include basal cell carcinoma and, to a lesser extent, squamous cell carcinoma, sebaceous carcinoma, porocarcinoma, and apocrine carcinoma. The lesion is engaging because its morphologic appearances vary with the age of the lesion.[3] Though it's technically classified as a hair follicle tumour and is associated with other conditions, a nevus sebaceous is benign. Here, we present a case of nevus sebaceous in a pediatric patient to highlight its clinical features, clinical examination, clinical diagnosis, management and histopathological examination.[4]

CASE PRESENTATION

A 12-year-old female patient came with chief complaint of painless swelling over the tip of nose. The swelling had been present for four years, during

which the parents observed progressive thickening and an uneven texture of the nasal skin. The lesion remained asymptomatic, with no associated itching, bleeding, pain, ulceration, or abnormal discharge. There was no reported family history of similar conditions.

Figure 1: Preoperative image

Figure 2: Postoperative image

A well-demarcated, slightly raised swelling was observed on the tip of the nose, measuring approximately 3 cm x 3 cm. The surface of the swelling was normal, with no evidence of scaling, crusting, or ulceration. The surrounding skin appeared normal in colour, texture, and temperature, with no signs of inflammation or erythema. The swelling was non-tender, firm in consistency, and non-pulsatile on palpation, with no fluctuation or warmth. No associated lymphadenopathy was detected in the cervical, submandibular, or preauricular regions. There were no signs of systemic involvement, and the patient no accompanying symptoms such as fever, weight loss, or malaise. Clinical diagnosis of Lipoma of the tip of nose was made.

Given the risk of neoplastic transformation, complete excision of the lesion was recommended. The lesion was excised under general anaesthesia, and the defect was closed with primary sutures. The excised mass was sent for histo-pathological examination. Postoperatively, the patient recovered uneventfully. Regular follow-up was advised for regular, meticulous dressing and monitoring for recurrence. Histopathological report showed keratinized stratified squamous epithelium with focal acanthosis and slight papillomatosis. The benign proliferation of sebaceous glands below the dermis extends to the reticular dermis without any granuloma, atypia, or malignant changes. There was also entrapment of mature adipocytes in stroma. On the basis of Histopathological findings, diagnosis of Nevus Sebaceous was made.

DISCUSSION

Nevus sebaceous, also known as organoid nevus, Jadassohn nevus, or pilosyringosebaceous nevus, is characterized as a rare, non-hereditary, congenital hamartoma, resulting from hyperplasia of epithelial, sebaceous, follicular, and apocrine elements of the skin.[5-7]

The etiology of this disease has not yet been fully clarified and, therefore, needs further investigation. However, recent studies suggest the association of a post-zygotic somatic mutation related to the HRAS (chromosome 11p15), NRAS (chromosome 1p13) and KRAS (chromosome 12p12) genes in the genesis of this condition, as they condition the cell proliferation process. The clinical manifestation occurs as plaques with partial or complete alopecia, with a linear or oval shape, and color ranging from skin-colored, to yellowish-orange or brownish-black, with a smooth, nipple-like or verrucous appearance, depending on the degree of lesion development.[8-10]

A 12-year-old female patient came with chief complaint of painless swelling over the tip of nose. The swelling had been present for four years, during which the parents observed progressive thickening and an uneven texture of the nasal skin. The lesion remained asymptomatic, with no associated itching, bleeding, pain, ulceration, or abnormal discharge. There was no reported family history of similar conditions. A well-demarcated, slightly raised swelling was observed on the tip of the nose, measuring approximately 3 cm x 3 cm. Clinical diagnosis of Lipoma of the tip of nose was made. Histopathological report showed keratinized stratified squamous epithelium with focal acanthosis and slight papillomatosis. The benign proliferation of sebaceous glands below the dermis extends to the reticular dermis without any granuloma, atypia, or malignant changes. There was also entrapment of mature adipocytes in stroma. On the basis of Histopathological findings, diagnosis of Nevus Sebaceous was made. Neto MPDS et al described the clinical manifestation of nevus sebaceous, as well as the main management techniques addressed in the medical literature. Nevus sebaceous occurs as lesions with a linear or oval appearance, with a smooth or verrucous texture, generally alopecic and with very variable color. Moreover, nevus

sebaceous is one of the components of the so-called linear nevus syndrome or Schimmelpenning-Feuerstein-Mims syndrome, which is associated with multisystemic complications.[10] The diagnosis of NSJ is usually clinical, and the follow-up and management of these lesions are unclear due to the risk of developing malignancies. Therefore, some authors recommend closer monitoring and excision only for aesthetic purposes or in cases where malignant change is suspected, rather than prophylactic surgery in late childhood or adolescence.[11]

In 1975, Solomon et al. analyzed the linear nevus sebaceous syndromes and their concomitant systemic disorders classifying them into one category under the common name of epidermal nevus syndrome. A revision of the previous division was presented by Happle, with a suggestion to classify the epidermal nevus syndromes into two groups. Schimmelpenning-Feuerstein-Mims syndrome, phacomatosis pigmentokeratotica, nevus comedonicus syndrome, angora hair nevus syndrome, and Becker nevus syndrome, which feature the presence of organoid epidermal nevi were categorized into well-defined phenotypes. This category also comprises the syndromes characterized by keratinocytic nevi, including CHILD syndrome (congenital hemidysplasia with ichthyosiform nevus and limb defects), type 2 segmental Cowden disease, Proteus syndrome and fibroblast growth factor receptor 3 epidermal nevus syndrome (García-Hafner-Happle syndrome).[12] The group of less well-defined phenotypes associated with epidermal nevi includes nevus trichilemmocysticus syndrome, didymosis aplasticosebacea, SCALP syndrome (sebaceous nevus, central nervous system malformations, aplasia cutis congenita, limbal dermoid and pigmented nevus), Gobello syndrome, Bäfverstedt syndrome, NEVADA syndrome (nevus epidermicus verrucosus with angiodysplasia and aneurysms), and CLOVE syndrome (congenital lipomatous overgrowth, vascular malformations, and epidermal nevus).[13] Kamyab-Hesari K et al reviewed the histopathologic features of 168 patients with nevus sebaceus to determine the frequency of different histologic features as well as associated neoplastic growths. Acanthosis, papillomatosis, and basal layer pigmentation were the most frequent findings. Hyperplastic changes of the sebaceous glands became obvious during the second decade and were present in 60% of the specimens from individuals older than 20. Inflammatory infiltrate was almost invariably present. Skin adnexal changes were frequently noted, including sebaceous gland hyperplasia (93.5%) and primitive hair follicles (76.8%). In 88.5% of scalp lesions, markedly decreased terminal hair was observed. Ectopic apocrine glands were present in 55.4% of specimens, while 24.4% showed anomalous ductal sweat gland structures resembling eccrine hyperplasia. Neoplastic changes were detected in nine cases, including four trichoblastomas, three tricholemmomas, and two syringocystadenoma papilliferums. Development of malignancies in sebaceus nevi is a rare phenomenon, and decision for excision of the lesion should be made after thorough evaluation of the pros and cons.[14]

CONCLUSION

Nevus sebaceous is a benign but dynamic lesion with potential for neoplastic transformation. Early identification and appropriate management, including surgical excision, are essential to prevent complications. This case supports the importance of routine monitoring and histopathological evaluation in managing nevus sebaceous.

REFERENCES

1. Lobato-Berezo A, Aguilera-Peiró P, Pujol-Vallverdú RM. Tumor Collision Over Sebaceous Nevus: Clues for Dermoscopic Diagnosis. Actas Dermosifiliogr (Engl Ed). 2018 Sep;109(7):647-648.
2. Sathyaki D.C., Riyas M., Roy M.S., Swarup R.J., Raghu N. Pigmented trichoblastoma of nose: an unusual occurrence. J Clin Diagn Res. 2017; 11:09–10.
3. El Ezzi O, de Buys Roessingh AS, Bigorre M, Captier G. Syndromic sebaceous nevus: current findings. Int J Dermatol. 2018 May;57(5):599-604.
4. Osman M.A.R., Kassab A.N. Carbon dioxide laser versus erbium: YAG laser in treatment of epidermal verrucous nevus: a comparative randomized clinical study. J Dermatolog Treat. 2017; 28:452–457.
5. Garcias-Ladaria J, Cuadrado Rosón M, Pascual-López M. Epidermal Nevi and Related Syndromes - Part 2: Nevi Derived from Adnexal Structures. Actas Dermosifiliogr (Engl Ed). 2018 Oct;109(8):687-698.
6. Wali G.N., Felton S.J., McPherson T. Management of naevus sebaceous: a national survey of UK dermatologists and plastic surgeons. Clin Exp Dermatol. 2018; 43:589–591.
7. Moreno-Arrones O.M., Perez-Garcia B. Nevus sebaceus on the face: experience with photodynamic therapy in adults and children. Indian J Dermatol Venereol Leprol. 2019; 85:440.
8. Pradhan S., Xiao H., Yang H.L., Ran Y.P. Dermoscopic clues for sebaceous carcinoma arising in nevus sebaceous. Chin Med J. 2020; 133:2121–2122.
9. Asch S, Sugarman JL. Epidermal nevus syndromes. Handb Clin Neurol. 2015; 132:291-316.
10. Neto MPDS, Assis BR, Andrade GR. Sebaceous nevus of Jadassohn: review and clinical-surgical approach. An Bras Dermatol. 2022 Sep-Oct;97(5):628-636.
11. Voiculescu VM, Celarel AM, Cozma EC, Banciu ML, Lupu M. Nevus Sebaceous of Jadassohn in Adults-Can Reflectance Confocal Microscopy Detect Malignant Transformation? Diagnostics (Basel). 2023 Apr 20;13(8):1480.
12. Happle R. The group of epidermal nevus syndromes Part I. Well defined phenotypes. J Am Acad Dermatol. 2010; 63:1–22.
13. Happle R, The group of epidermal nevus syndromes Part II. less well defined phenotypes. J Am Acad Dermatol. 2010; 63:25–30.
14. Kamyab-Hesari K, Seirafi H, Jahan S, Aghazadeh N, Hejazi P, Azizpour A, Goodarzi A. Nevus sebaceus: a clinicopathological study of 168 cases and review of the literature. Int J Dermatol. 2016 Feb;55(2):193-200.