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Case Presentation

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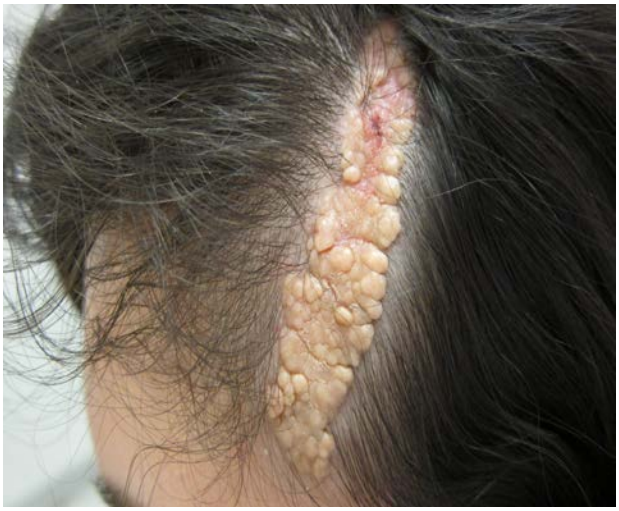
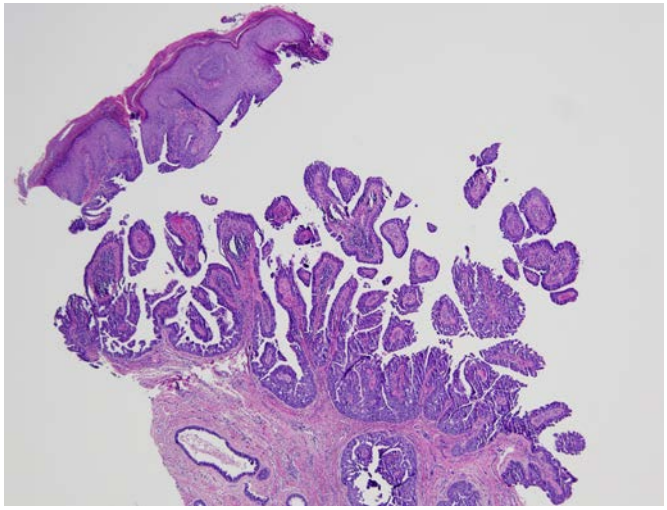
Abstract

We present a case of a 35-year-old woman with a yellow, verrucous, and itchy plaque on her scalp. Within this plaque, there was an erythematous, bleeding papule. Histopathologic findings were compatible with a diagnosis of syringocystadenoma papilliferum within a nevus sebaceous. We present a brief review of the natural history of nevus sebaceus, its pathogenesis, and management.





Case synopsis

A 35-year-old woman presented to the Dermatology Clinic at Bellevue Hospital Center with a pruritic plaque on her head that has been present since birth. There is now an area within the lesion that bleeds. She is interested in having the lesion removed.

Physical Examination: On the left frontal scalp there was a large, yellow, verrucous plaque with alopecia. At the superior aspect there was a red papule with central pinpoint bleeding.

Histopathology: Emanating from a hyperplastic epidermis, there are papillations that are lined by bi-layered cuboidal epithelium with apocrine differentiation and a dense, lymphoplasmacytic inflammatory infiltrate.

Diagnosis: Syringocystadenoma papilliferum in a nevus sebaceus

Discussion: Nevus sebaceus is a benign, congenital, cutaneous hamartoma with epidermal, sebaceous, and apocrine components with an incidence of 1 in 1000 live births. It presents as an alopecic, waxy, yellowish plaque, which is commonly found on the scalp and face. Histopathologic features include epithelial hyperplasia, prominent mature sebaceous glands, and poorly formed hair follicles [1]. Ectopic apocrine glands may be observed focally [2].

The natural history of nevus sebaceus may be divided into three stages. Early in life, the lesion is characterized by papillomatous hyperplasia and the presence of immature hair follicles. The second stage occurs at the time of puberty. Hormonal changes result in the development of the sebaceous glands and apocrine glands. The lesion clinically appears as a verrucous plaque. In the third stage, various benign and malignant neoplasms may occur within the lesion [3].

In a retrospective analysis of 596 surgical excision specimens of nevus sebaceus, more than 90% of the associated tumors were benign. The three most common tumors were syringocystadenoma papilliferum, trichoblastoma, and trichilemmoma [4]. In a meta-analysis of 4923 cases of nevus sebaceus, the most common malignant tumors observed were basal-cell carcinoma, sebaceous carcinoma, and squamous-cell carcinoma [3]. Although most tumors develop in the third stage of evolution of the lesion, malignant transformation may occur even before puberty and without any notable clinical change in the lesion [5].

Syringocystadenoma papilliferum is a benign neoplasm of apocrine differentiation with a tubular and/or papillary microscopic pattern. Most papillary syringocystadenomas that present on the scalp occur in association with a nevus sebaceus. Syringocystadenoma papilliferum clinically appears as grouped papules and nodules with scale-crust and serosanguinous drainage. This neoplasm consists of papillary foci that are lined by columnar cells with apocrine differentiation. The dermal cores of the papillae usually are filled with lymphocytes and plasma cells. The papillary fronds are in continuity with the surface squamous epithelium [6].

On dermoscopic examination of syringocystadenoma papilliferum within a nevus sebaceus, the lesion may appear as an erythematous papule or nodule, which is divided by white linear structures that define variably sized lobules and contain different vascular structures, such as irregular linear vessels, glomerular vessels, and vessels in a horseshoe arrangement [7].

In a recent evaluation of 65 specimens of nevus sebaceus, cutaneous mosaicism for postzygotic activating mutations of HRAS and KRAS were found to cause nevus sebaceus. The HRAS c.37G>C mutation, which results in a p.Gly13Arg substitution, was noted in 91% of lesions. Functional analysis of the HRAS c.37G>C confirmed constitutive activation of the MAPK and PI3K-Akt signaling pathways. This mutation was also found in eight of eight secondary tumors, which included syringocystadenoma papilliferum, trichoblastomas, and trichilemmomas [1].

Once a malignant lesion arises within a nevus sebaceus, the general consensus is that the tumor and entire nevus should be excised. Excisions should be full-thickness, through the epidermis, dermis, and subcutaneous tissue, with 2-to-3mm margins [3]. Mohs micrographic surgery is an alternative treatment option [8]. Prophylactic excision, however, is controversial. The unknown risk of malignant transformation, increased lesion friability, associated bleeding, itching, aesthetic concerns associated with scalp alopecia, and lesion expansion might prompt early excision. However, these concerns need to be weighed against the risks of general anesthesia in children and the risks of excision, such as bleeding, infection, and scarring. Other destructive modalities, such as electrosurgery, dermabrasion, carbon-dioxide laser ablation, and photodynamic therapy, are not effective in removing the entire lesion and may mask the appearance of malignant conditions [3].

References

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