Case Reports in Dermatology

Case Rep Dermatol 2016;8:75–79

DOI: 10.1159/000445822
Published online: April 20, 2016

1662–6567/16/0081–0075$39.50/0
www.karger.com/cde

Four Different Tumors Arising in a Nevus Sebaceous

Takeshi Namiki[a] Keiko Miura[b] Makiko Ueno[a] Yumi Arima[a]
Aya Nishizawa[a] Hiroo Yokozeki[a]

Departments of [a]Dermatology and [b]Pathology, Graduate School of Medical and Dental Sciences, Tokyo Medical and Dental University, Tokyo, Japan

Key Words
Syringocystadenoma papilliferum · Sebaceoma · Trichoblastoma · Basal cell carcinoma · Nevus sebaceous of Jadassohn · Multiple tumors

Abstract
Nevus sebaceous is known by its association with one or more secondary tumors, but more than three multiple tumors arising from a nevus sebaceous is extremely rare. A 67-year-old female presented with a light brown plaque on the back of her head that contained a dome-shaped black node and an erosive lesion. Histopathological examination showed atypical basaloid cells in the black node. At the periphery of that node, structures resembling follicular germs extruded from interlacing cords in the upper portion and tumor nests with sebocytes were in the lower portion. In the erosive lesion, papillated structures with an apocrine epithelium were observed. In the light brown plaque, enlargement of sebaceous lobules was noted. From those histopathological features, a diagnosis of syringocystadenoma papilliferum, sebaceoma, trichoblastoma and basal cell carcinoma arising from a nevus sebaceous was made. We discuss the rarity of multiple tumors arising from a nevus sebaceous.

Introduction

Nevus sebaceous of Jadassohn is a congenital hamartoma, which has been reported to be associated with secondary tumors such as syringocystadenoma papilliferum, trichoblastoma and basal cell carcinoma (BCC) [1–4]. However, more than three multiple tumors arising from a nevus sebaceous of Jadassohn is extremely rare [5]. Malignant transformation occurs in 10–15% of nevus sebaceous in some series, although others suggest that this rate

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Takeshi Namiki, MD, PhD
Department of Dermatology, Graduate School of Medical and Dental Sciences
Tokyo Medical and Dental University
1-5-45, Yushima, Bunkyo-ku, Tokyo 113-8510 (Japan)
E-Mail tnamderm@tmd.ac.jp

may be lower. The most common malignant neoplasm arising in this disorder is BCC. Studies indicate that the development of BCC or any other malignant neoplasm is very rare. The most common tumors arising within nevus sebaceous are syringocystadenoma papilliferum and trichoblastoma. Multiple tumors arising from a nevus and/or a phacomatosis provide important clues to understand their histogenesis and their capacity to differentiate and proliferate. Here we present a case of multiple tumors of syringocystadenoma papilliferum, sebaceoma, trichoblastoma and BCC that arose from a nevus sebaceous of Jadassohn.

Case Report

A 67-year-old female presented with a tumor on the back of her head. The patient had recognized a nodule on the back of her head 37 years earlier. That nodule had gradually enlarged and had started bleeding 10 years earlier. Physical examination revealed a 47 × 26 mm slightly elevated light brown plaque on the back of her head. A 16 × 13 mm erosive lesion and an 18 × 17 mm dome-shaped black node were recognized in that plaque (fig. 1a). Dermoscopic analysis of the dome-shaped black node showed black homogenous areas, surface scales and arborizing vessels (fig. 1b). Dermoscopic analysis of the erosive lesion showed glomerular vessels in pink homogenous areas (fig. 1c).

Histopathological examination of the black node showed tumor nests with a sheet-like appearance that extended downward into the deep dermis. Melanin deposition was scattered irregularly and bleeding was also observed within the tumor nests (fig. 2a). The tumor cells were atypical and basaloid, and heterotypic mitoses were observed. Insular tumor aggregations were recognized in the tumor nests and some of those aggregations contained melanin deposits (fig. 2b). Immunohistochemistry showed a positive reaction for Ber-EP4 in those tumor nests (fig. 2c). We made a diagnosis of BCC at this portion. At the periphery of the black node, tumor nests also extended into the deep dermis and were composed of several different components (fig. 2d). In the upper portion, tumor nests with interlacing cords were noted. Some of those tumor nests exhibited cribriform and reticular patterns (fig. 2e). Numerous structures resembling follicular germinative cells were recognized and some of those structures extruded from interlacing cords (fig. 2f). A diagnosis of trichoblastoma was made in this upper portion. In the lower portion, tumor nests exhibited lobular aggregations, and had ductal and/or cystic structures (fig. 2g). The tumor nests consisted of sebocytes, which had a foamy cytoplasm (fig. 2h). Immunohistochemistry showed a positive reaction for adipophilin in those tumor nests (fig. 2i). From those histopathological findings, a diagnosis of sebaceoma was made. In the erosive lesion, papillated structures with apocrine epithelium were observed. Tubules lined by apocrine epithelium had an anastomosing pattern (fig. 2j). The epithelium of the tubules was lined by 2–3 layers displaying apocrine secretion of decapitation secretion and numerous plasma cells were noted in the stroma of the papillated structures (fig. 2k). A diagnosis of syringocystadenoma papilliferum was made at the erosive lesion. In the slightly elevated light brown plaque, the epidermis was hyperkeratotic and had a slightly anastomosing structure. Enlargement of sebaceous lobules connecting with abnormal follicular infundibula was noted (fig. 2l). From those histopathological features, a diagnosis of syringocystadenoma papilliferum, sebaceoma, trichoblastoma and BCC arising from a nevus sebaceous of Jadassohn was made. The tumor was resected and there has been no relapse 1 year after the surgery.

Discussion

Nevus sebaceous has the potential to generate different lineages of tumors that are not restricted to the sebaceous lineage [1, 4]. A secondary tumor from a nevus sebaceous may differentiate into follicular, sebaceous, apocrine and eccrine cells, and rarely, a secondary tumor can differentiate into muscle [6, 7]. This fact suggests the pluripotency of nevus sebaceous. The simultaneous occurrence of tumors from a nevus sebaceous is not rare, but the number of tumors is usually less than three. The incidence of more than three multiple tumors is quite unusual [5]. A previous study showed that one in 140 cases of nevus sebaceous simultaneously developed three tumors, and another previous study showed that one in 150 cases of nevus sebaceous simultaneously developed four tumors [8, 9]. In our case, four distinct tumors (syringocystadenoma papilliferum, sebaceoma, trichoblastoma and BCC) arose from a nevus sebaceous. Those tumors reflect that nevus sebaceous has a pluripotency for apocrine, sebaceous and follicular differentiation. An intriguing fact is that this patient had a previous history of multiple tumors such as ovarian cystoma, submandibular gland tumor and thyroid cancer.

In conclusion, we report a rare case of multiple tumors of syringocystadenoma papilliferum, sebaceoma, trichoblastoma and BCC arising from a nevus sebaceous of Jadassohn. The simultaneous development of four multiple tumors from a nevus sebaceous is extremely rare.

Statement of Ethics

The patient gave written informed consent

Disclosure Statement

The authors declare no conflicts of interest.

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Fig. 1. **a** A 47 × 26 mm slightly elevated light brown plaque on the back of the patient's head. A 16 × 13 mm erosive lesion and an 18 × 17 mm dome-shaped black node were recognized in this plaque lesion. The left dotted line indicates the histopathological sections shown in fig. 2a–c, the center dotted line indicates the sections shown in fig. 2d–i, and the right dotted line indicates the sections shown in fig. 2j–l. **b** Dermoscopic features of the dome-shaped black node showing black homogenous areas, surface scales and arborizing vessels. **c** Dermoscopic features of the erosive lesion showing glomerular vessels in pink homogenous areas.

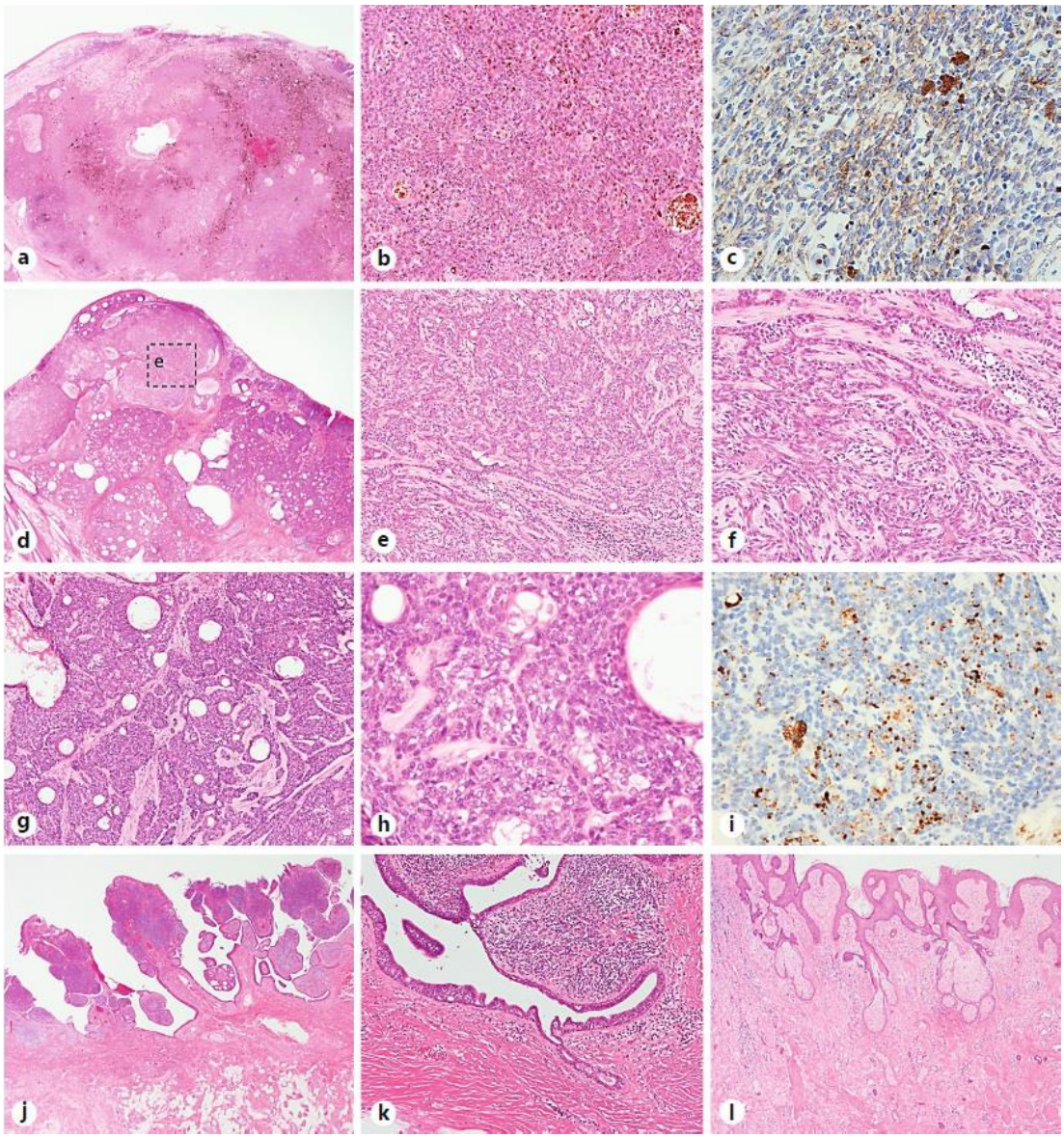

Fig. 2. **a** Histopathological findings from the black node showing tumor nests with a sheet-like appearance extending downward into the deep dermis and melanin deposits scattered irregularly (HE stain; original magnification, ×20). **b** Each tumor cell was atypical and insular tumor aggregations were recognized in the tumor nests (HE stain; original magnification, ×200). **c** Immunohistochemistry for Ber-EP4 (original magnification, ×400). **d** Histopathological findings at the periphery of the black node showing tumor nests composed of several different components. The dotted box indicates the portion shown in **e** (HE stain; original magnification, ×12.5). **e** In the upper portion, tumor nests with interlacing cords were noted (HE stain; original magnification, ×100). **f** Numerous structures resembling follicular germinative cells were recognized and some of those structures extruded from interlacing cords (HE stain; original magnification, ×200). **g** In the lower portion, the tumor nests exhibited lobular aggregations and had ductal and/or cystic structures (HE stain; original magnification, ×100). **h** Tumor nests consisting of sebocytes with a foamy cytoplasm (HE stain; original magnification, ×400). **i** Immunohistochemistry for adipophilin (original magnification, ×400). **j** In the erosive lesion, papillated structures with apocrine epithelium were observed (HE stain; original magnification, ×20). **k** The epithelium of tubules displayed decapitation secretion and numerous plasma cells were noted in the stroma (HE stain; original magnification, ×100). **l** In the slightly elevated light brown plaque, the epidermis was hyperkeratotic and enlargement of sebaceous lobules was noted (HE stain; original magnification, ×40).