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# Nevus sebaceus with syringocystadenoma papilliferum, prurigo nodularis, apocrine cystadenoma, basaloid follicular proliferation, and sebaceoma: case report and review of nevus sebaceus-associated conditions

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## Abstract

Nevus sebaceus is a benign skin hamartoma of congenital onset that grows during puberty, and in adulthood can develop secondary benign and malignant neoplasms. The most common benign neoplasms occurring in nevus sebaceus are believed to be syringocystadenoma papilliferum, trichilemmoma, and trichoblastoma. A patient with nevus sebaceus developed not only syringocystadenoma papilliferum but also prurigo nodularis within her hamartomatous lesion; multiple biopsies were necessary to establish the diagnoses. Excision of the residual nevus sebaceus also revealed an apocrine cystadenoma, basaloid follicular proliferation, and sebaceoma. Also, it is important to select the appropriate biopsy site and size when evaluating a patient for secondary neoplasms within their nevus sebaceus. Indeed, more than one biopsy may be required if additional diagnoses are suspected.

*Keywords: benign, neoplasm, nevus, nodularis, papilliferum, prurigo, sebaceus, secondary, syringocystadenoma, tumor*

## Introduction

Nevus sebaceus, also known as nevus sebaceus of Jadassohn or organoid nevus, is a rare congenital benign skin hamartoma [1]. Lesions typically appear at birth and grow during puberty; in adulthood, they may develop secondary neoplasms [2-5]. A woman

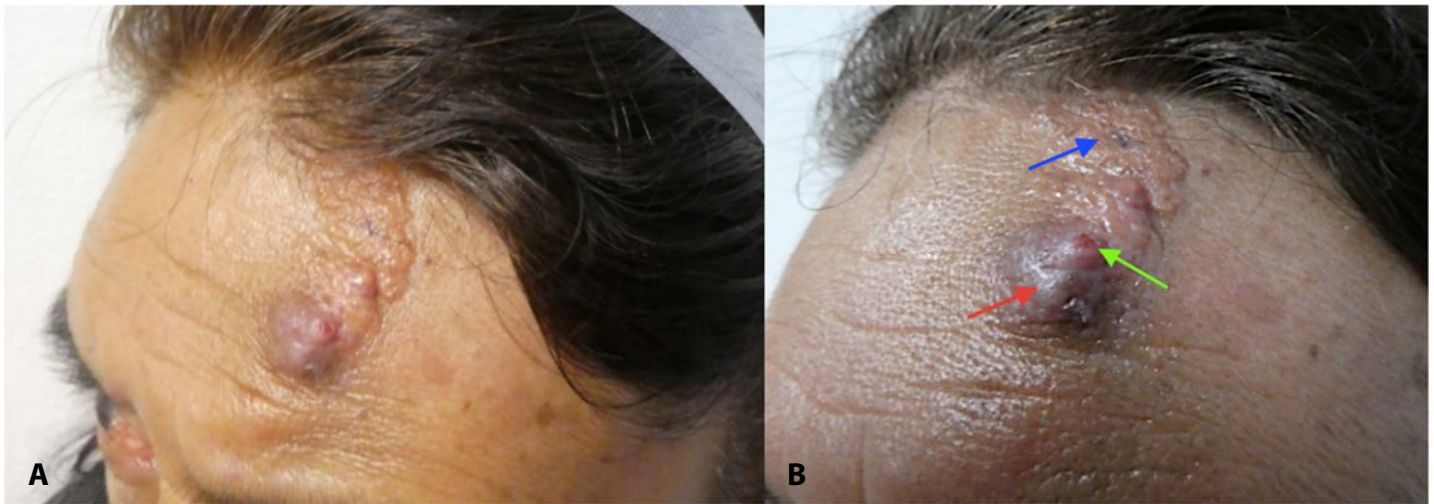
with nevus sebaceus who not only developed syringocystadenoma papilliferum but also prurigo nodularis in the inferior portion of her lesion is described. Complete excision of the residual nevus sebaceus also revealed three concurrent additional conditions: apocrine cystadenoma, basaloid follicular proliferation, and sebaceoma. The literature of nevus sebaceus-associated conditions is reviewed.

## Case Synopsis

A healthy 67-year-old woman with a history of gastroesophageal reflux disease on metoclopramide presented for evaluation of a lesion on her forehead that had been present since birth. It became slightly thicker as a teenager after puberty and remained unchanged until three years prior to presentation. At that time, the lower portion of the plaque not only began to increase in size but also started to itch. On further questioning, she mentioned that she chronically rubbed the area.

Cutaneous examination revealed a 6.5 by 1.5-centimeter flesh-colored plaque on the left side of her forehead. In the inferior portion of the lesion, there was a purple 15 by 12-millimeter firm nodule. Two millimeter-sized punch biopsies were performed of the superior plaque portion and a papular area in the nodular portion (**Figure 1**).

Microscopic examination of the specimen from the plaque portion revealed hyperkeratosis, acanthosis,



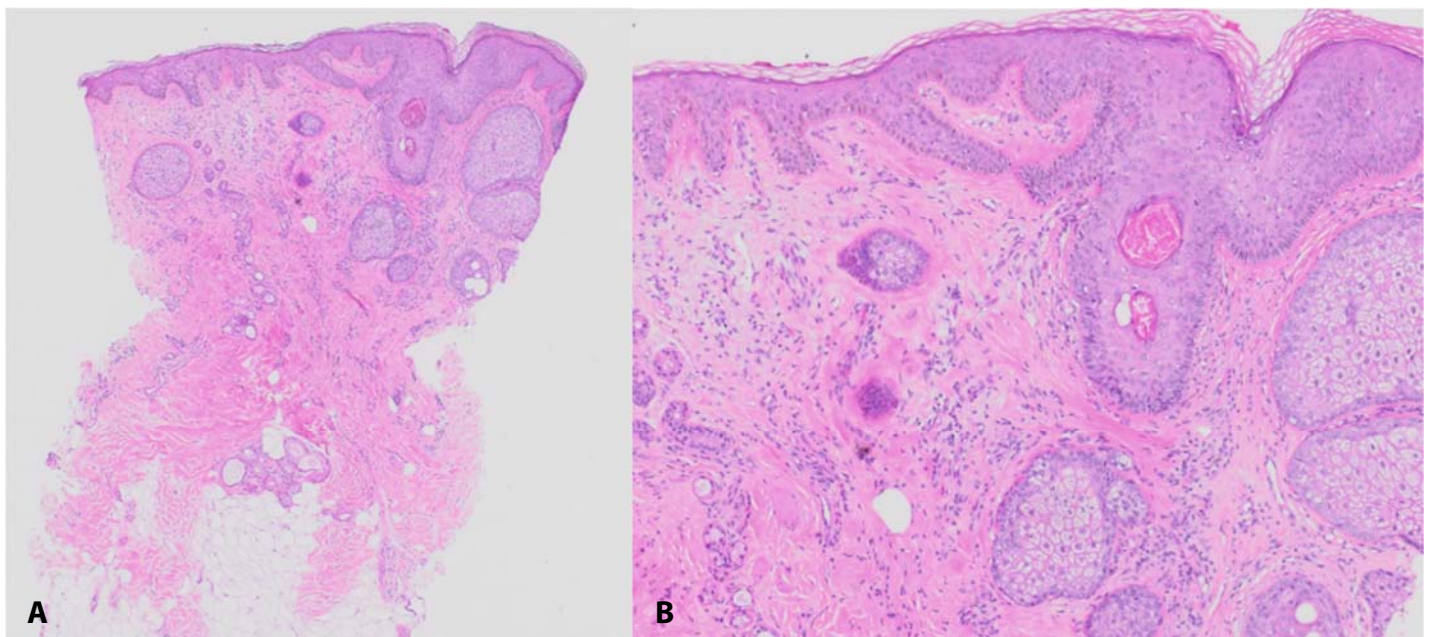
**Figure 1.** Nevus sebaceus with secondary syringocystadenoma papilliferum and prurigo nodularis. **A)** Distant and **B)** close-up views of a 67-year old woman with a flesh-colored plaque on her forehead consistent with a nevus sebaceus. The superior biopsy site (blue arrow) and inferior biopsy site (red arrow) from her initial visit showed nevus sebaceus and prurigo nodularis, respectively. A second skin biopsy (green arrow) of the new purple nodule revealed a syringocystadenoma papilliferum.

and mild papillomatosis. In the dermis, there were numerous sebaceous glands; in addition, apocrine glands were present in the deep reticular dermis. These findings established the diagnosis of nevus sebaceus (**Figure 2**).

Microscopic examination of the papule showed hyperkeratosis and irregular acanthosis. In addition, there was thickening of the collagen bundles in the

papillary dermis. These findings established the diagnosis of prurigo nodularis (**Figure 3**).

Although the patient confirmed that she rubbed the itchy inferior portion of her lesion, the diagnosis of prurigo nodularis did not seem to entirely account for the new tumor that had developed. Therefore, given the questionable clinical correlation and the small size of the biopsy relative to the size of the



**Figure 2.** Pathology features of nevus sebaceus. **A)** Distant, and **B)** closer magnification views of hematoxylin and eosin stained sections of a nevus sebaceus on the forehead of a 67-year-old woman. The epidermis shows hyperkeratosis, acanthosis, and mild papillomatosis. There are several sebaceous glands; apocrine glands are also present in the deep reticular dermis. **A)** 2 $\times$ ; **B)** 10 $\times$ .

lesion, another biopsy, a four millimeter-sized punch biopsy, was performed at the new inferior aspect of the tumor.

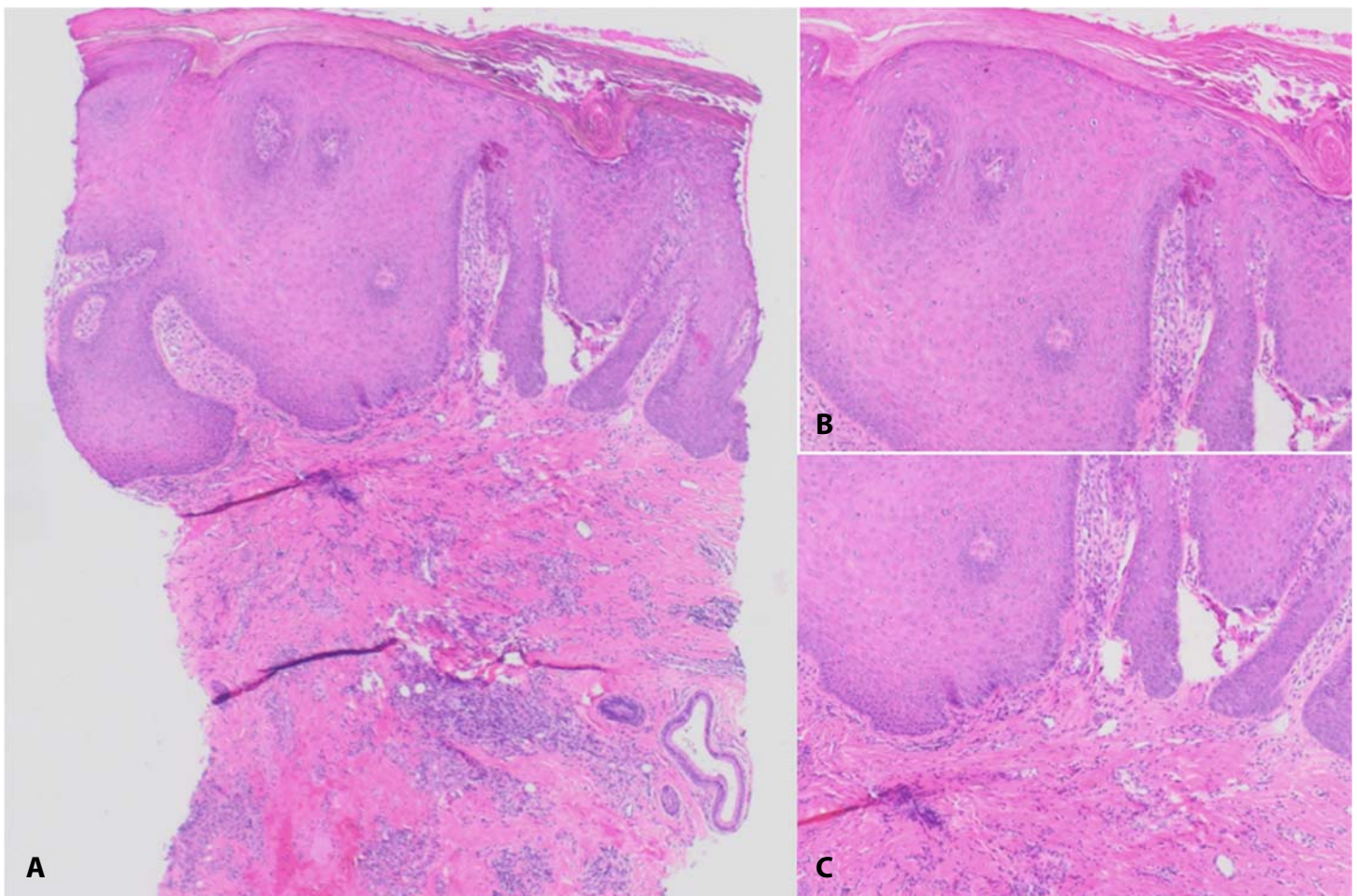
The second biopsy of the new inferior nodule revealed a benign adnexal neoplasm with papillomatous apocrine differentiation. The inflammatory infiltrate in the dermal stroma consisted of plasma cells. These findings established the diagnosis of a syringocystadenoma papilliferum (**Figure 4**).

Correlation of the patient's clinical presentation and pathology findings demonstrated that she had a nevus sebaceus with subsequent development of not only a syringocystadenoma papilliferum within the original tumor, but also prurigo nodularis secondary to chronic rubbing. The residual nevus sebaceus was completely excised. Microscopic

examination of the specimen also showed an apocrine, a benign basaloid follicular proliferation and a sebaceoma.

### Case Discussion

Nevus sebaceus is a rare congenital hamartoma of the epidermis, dermis, and appendages with a prevalence of sebaceous elements [1]. Nevus sebaceus occurs in approximately 0.3 percent of newborns without sex predilection [6]. During puberty, these lesions undergo a growth phase that is thought to be hormonally driven. In adulthood, the growths may develop secondary neoplasms within them, a phenomenon thought to occur in about 20 percent of cases and typically after the age of forty [4].



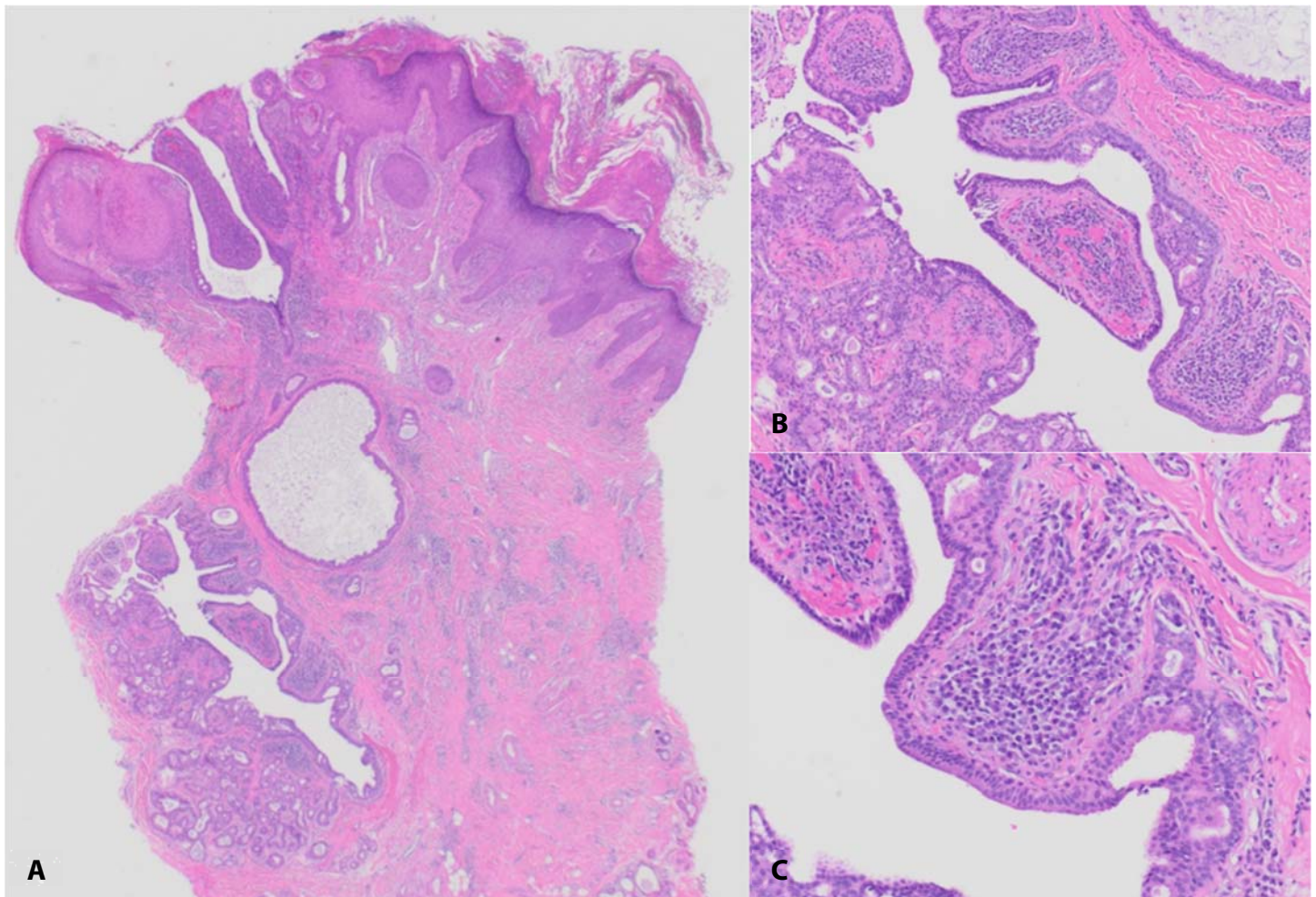
**Figure 3.** Pathology features of prurigo nodularis. **A)** Distant and **B, C)** closer magnification views of hematoxylin and eosin stained sections of prurigo nodularis from the forehead of a 67-year-old woman. There is hyperkeratosis and irregular acanthosis of the epidermis. There are thickened collagen bundles in the papillary dermis (hematoxylin and eosin: **A)** 4x; **B)** 10x; **C)** 10x.

Nevus sebaceus lesions appear at birth as waxy, yellow-orange or tan, hairless plaques commonly on the scalp with a localized area of alopecia [3]. However, they may also occur on the forehead, face, or neck [2, 7]. Lesions range in size from one to several centimeters. Recent etiopathogenic studies suggest an association with human papillomavirus or patched gene mutations [4].

Histologically, nevus sebaceus is associated with thickening of the epidermis with varying degrees of acanthosis and papillomatosis. In early lesions, sebaceous glands may be underdeveloped and decreased in number. Immature hair follicles are characteristic and diagnostic. After puberty, the epidermis shows prominent papillomatous

hyperplasia and there is an increased number of sebaceous and apocrine glands [3, 8].

Benign and malignant epithelial neoplasms associated with nevus sebaceus are summarized in **Table 1** [2, 5, 8]. Previously, the most common tumor identified was basal cell carcinoma. However, subsequent studies have suggested that basal cell carcinomas were misdiagnosed and that nevus sebaceus is actually associated with trichoblastoma, a benign hair follicle tumor, whose pathologic features may mimic those of basal cell carcinoma [8]. Other common tumors which may emerge include syringocystadenoma papilliferum and trichilemmoma [5, 9]. The prurigo nodularis present in our patient's tumor is postulated to have resulted



**Figure 4.** Pathology features of syringocystadenoma papilliferum. **A)** Distant and **B, C)** closer magnification views of hematoxylin and eosin stained sections of a syringocystadenoma papilliferum from the forehead of a 67-year-old woman. There is a tumor with papillomatous apocrine differentiation. There are papillomatous and cystic invaginations lined by apocrine glandular cells extending from the epidermis into the dermis. There is an abundant plasma cell inflammatory infiltrate in the dermis hematoxylin and eosin: **A)** 4x; **B)** 10x; **C)** 20x.

**Table 1.** Secondary neoplasms associated with nevus sebaceus<sup>a</sup>.

Benign	Malignant
Apocrine cystadenoma	
Basaloid proliferation	
Ecrrine spiradenoma	
Hidradenoma	Adnexal carcinoma
Infundibuloma	Apocrine carcinoma
Osteoma	Basal cell carcinoma
Sebaceoma	Keratoacanthoma
Sebaceous adenoma	Mucoepidermoid carcinoma
Seborrheic keratosis	Sebaceous carcinoma
Syringocystadenoma papilliferum	Sebaceous epithelioma
Syringoma	Squamous cell carcinoma
Trichilemmoma	
Trichoblastoma	
Verruca vulgaris	

<sup>a</sup>Our patient, a 67-year-old Hispanic woman, had a nevus sebaceus associated with apocrine cystadenoma, benign basaloid follicular proliferation, sebaceoma, and syringocystadenoma papilliferum; in addition, prurigo nodularis was also present.

from recurrent rubbing of the lesion and does not represent a bona fide nevus sebaceus-associated neoplasm.

Approximately ten to 20 percent of nevus sebaceus is associated with a syringocystadenoma papilliferum [9]. Histologically, syringocystadenoma papilliferum is a benign adnexal tumor of apocrine derivation composed of papillary projections and cystic ducts lined by cuboidal or columnar cells with abundant basophilic cytoplasm and usually a connection to the overlying epidermis [10]. As observed in our patient, the dermal stroma characteristically contains plasma cells.

Our patient had five growths concurrently present in her nevus sebaceus: syringocystadenoma papilliferum, prurigo nodularis, apocrine cystadenoma, basaloid follicular proliferation, and sebaceoma. To the best of our knowledge, this is the first report of prurigo nodularis being described in

nevus sebaceus. Our patient's prurigo nodularis likely occurred secondary to pruritis of the lesion followed by chronic rubbing of the tumor.

The clinical course of our patient suggested the scenario of a secondary neoplasm developing in her nevus sebaceus. The initial biopsy showed a reactive epithelial process, yet the pathology diagnosis did not entirely correlate with the clinical presentation. Thus, a repeat biopsy was performed to establish the additional diagnosis; the diameter of the biopsy was also increased to ensure that an adequate sample was obtained. Unexpectedly, three additional conditions were also diagnosed after microscopic examination of the entire tumor following complete excision.

## Conclusion

Nevus sebaceus is a benign hamartoma of congenital onset, which increases in size at puberty and may develop subsequent secondary neoplasms. Our patient developed not only syringocystadenoma papilliferum but also prurigo nodularis, apocrine cystadenoma, basaloid follicular proliferation, and sebaceoma in her nevus sebaceus. Although syringocystadenoma papilliferum and trichoblastoma are the most common nevus sebaceus-associated secondary neoplasms, other benign and malignant neoplasms may develop in nevus sebaceus. Adequate biopsy size and appropriate site selection are necessary to ensure the correct diagnosis. When the pathology findings do not entirely correlate with the clinical presentation, an additional biopsy may be necessary to establish the diagnosis.

## Potential conflicts of interest

The authors declare no conflicts of interests.

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