Basal Cell Carcinoma Arising in a Port-Wine Stain

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BACKGROUND. **The occurrence of basal cell carcinoma within a port-wine stain or nevus flammeus is rare. Sixteen cases of basal cell carcinoma which developed in a port-wine stain or nevus flammeus have been reported.**

OBJECTIVE. **The objective was to demonstrate a rare case of basal cell carcinoma occurring in a port-wine stain successfully treated with Mohs micrographic surgery.**

METHODS. **This is a case report and literature review.**

RESULTS. **An 87-year-old man presented with a basal cell carcinoma on the margin of a previously untreated port-wine stain on the left cheek. Histologic examination showed a nodular basal cell carcinoma. The basal cell carcinoma was completely excised with Mohs micrographic surgery and complex linear closure was used to repair the wound in layers. The postoperative course was complicated by a hematoma, which developed 24 hr postoperatively. The hematoma was drained and there was no further bleeding or evidence of recurrence of the tumor after 12 months.**

CONCLUSION. **Basal cell carcinoma should be included in the differential diagnosis of a skin lesion occurring in a port-wine stain.**

SIRUNYA SILAPUNT, MD, LEONARD H. GOLDBERG, MD, FRCP, MARZIEH THURBER, MD, AND PAUL M. FRIEDMAN, MD HAVE INDICATED NO SIGNIFICANT INTEREST WITH COMMERCIAL SUPPORTERS.

PORT-WINE STAINS occur in 0.3% of neonates.[1] Port-wine stain may occur as an isolated skin lesion or be associated with other abnormalities such as Sturge-Weber syndrome (neural) and Klippel-Trenaunay-Weber syndrome (bones and soft tissues).[2] With advancing age, the color of port-wine stain gradually deepens/darkens and the surface thickens, becoming raised and nodular. The development of basal cell carcinoma in the skin damaged by prolonged exposure to sunlight and ionizing radiation has been well documented.[3,4]

The occurrence of basal cell carcinoma within a port-wine stain or nevus flammeus is rare. Sixteen cases of basal cell carcinoma which developed in a port-wine stain or nevus flammeus have been reported in the literature de novo or following the use of thorium X, Grenz ray or laser treatment (Table 1).

Case Report

An 87-year-old man presented with a 1.3×1.1-cm ulcerated nodule at the lower margin of his port-wine stain (Figure 1). The port-wine stain had grown steady in size and thickness over the lifetime of the patient. He had never had treatment of any kind for the port-wine stain. The medical history and underlying diseases in our patient included an atrial arrhythmia, mitral valve prolapse, bacterial endocarditis, stroke, mild heart failure, and prostate cancer. He was taking the following medications: angiotensin II inhibitor (Micardis), digitalis (Lanoxin), metoprolol (beta-blocker), potassium (K-Dur), diuretic (Demadex), and tamsulosin (Flomax). He was not on anticoagulants for his underlying cardiac disease or any herbals or supplements that served as anticoagulants. There was no history of bleeding with surgery or known bleeding disorder.

Examination of the skin revealed a large reddish purple port-wine stain with nodular surface on the left side of the face in the region innervated by the second division of the trigeminal nerve. A sclerosing and ulcerated nodule measuring 1.3×1.1 cm in diameter was found at the lower margin of the port-wine stain (Figure 2). The histologic examination showed a nodular basal cell carcinoma. The basal cell carcinoma was excised with Mohs micrographic surgery. Tumor-free margins were obtained after two stages of Mohs micrographic surgery. There was no excessive bleeding during surgery. The cautery machine was not needed and the bleeding was controlled by a pressure dressing alone. The surgical wound (4.2×3.8 cm in diameter) was repaired with a complex linear closure in layers which resulted in a 7-cm surgical scar placed horizontally along the lower margin of port-wine stain (Figures 3–5). Because the patient was older, he had abundant donor skin, which could be moved into

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Table 1. Previously Reported Cases of Basal Cell Carcinoma Arising in Port-Wine Stain or Nevus Flammeus

Authors	*Year*	*Age/Sex*	*Site*	*Type of Lesion*	*Therapy for Port-Wine Stain*	*Therapy for Basal Cell Carcinoma*	*No. of Basal Cell Carcinomas*
Scott[5]	1948	38/M	R hypochondrium	Nevus flammeus	None	Not mentioned	1
Rogers[6]	1954	10/M	R preauricular	Port-wine stain	Thorium X (35 applications, at ages 3–9)	Excision	1
Courtemanche[7]	1961	16/M	R lower eyelid and chin	Port-wine stain	Thorium X and carbon dioxide snow (at age 7)	Excision & grafting	1
Sarkarny and Caron[8]	1965	40/M	R temple	Port-wine stain	None	Excision & grafting	1
Sagi et al.[9]	1984	44/M	R side of face	Port-wine stain	Grenz ray (at age 6)	Excision & grafting	Multiple
Magana-Garcia and Magana-Lozano[10]	1988	49/M	L cheek and chin	Port-wine stain	None	Excision	12
Duhra and Foulds[11]	1991	63/M	L cheek	Port-wine stain	None	Excision & flap	1
Lo et al.[12]	1991	69/M	R forehead, R upper lip	Nevus flammeus	None	Mohs micrographic surgery	1
Shah et al.[13]	1996	47/F	R cheek	Port-wine stain	Thorium X (at early childhood)	Excision	2
		75/M	R cheek	Port-wine stain	Thorium X (during childhood)	Not mentioned	2
		44/F	R cheek	Port-wine stain	Thorium X (at ages 9 and 15)	Excision radiotherapy	2
Sarhadi and Soutar[14]	1997	59/F	Chin	Port-wine stain	Copper vapor laser	Excision	2
Wharton and Cole[15]	2001	41/M	R upper lip	Port-wine stain	Argon laser (at age 21)	Excision	1
		57/M	R cheek	Port-wine stain	Radium plates	Excision	5
		65/M	L cheek and upper lip	Port-wine stain	Radiotherapy	Excision & Grafting	1
		49/M	R neck	Port-wine stain	Topical radiotherapy (during childhood)	Excision	1

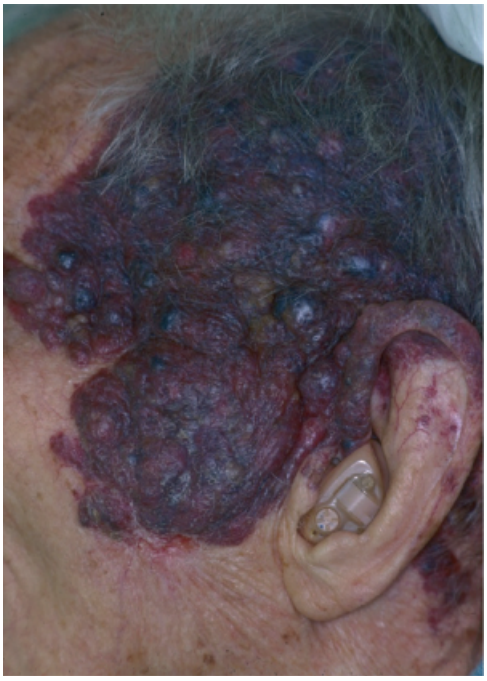

Figure 1. An ulcerated, sclerosing plaque on the inferior margin of a port-wine stain.

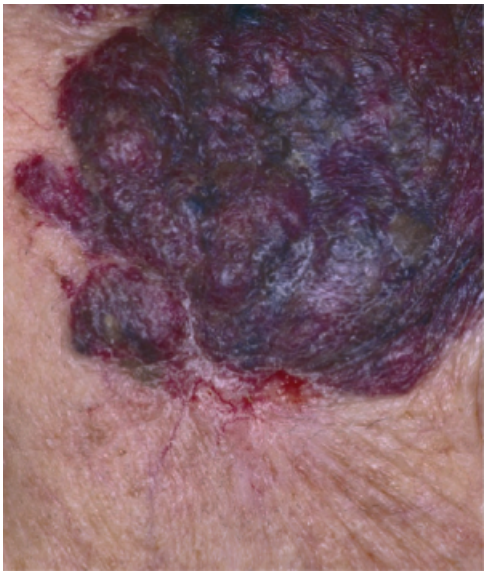

Figure 2. Close-up of Figure 1.

position to close the defect without undermining. Wound tension was moderate and was considered a positive feature of closure in that it provided pressure to support hemostasis.

The patient had antibiotic prophylaxis (500 mg amoxicillin three times a day) for the prevention of bacterial endocarditis beginning the day before the surgery and continuing for 7 days. The day following the Mohs micrographic surgery, the patient presented with

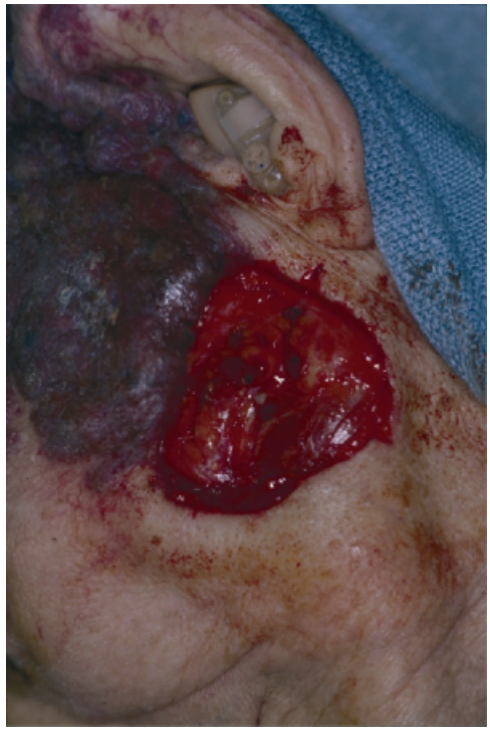

Figure 3. Postoperative defect.

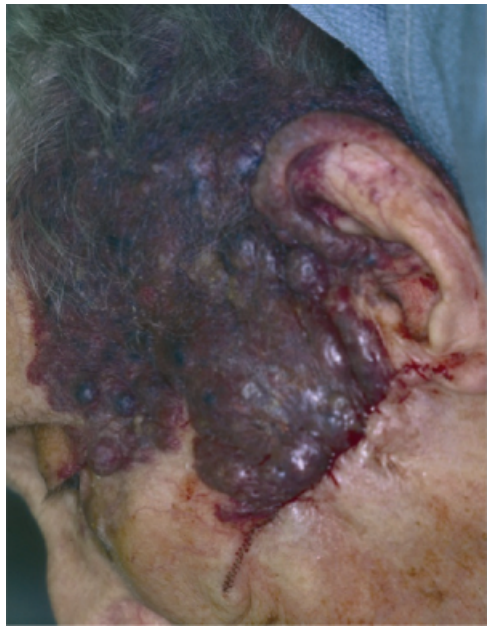

Figure 4. Postoperative closure.

a hematoma at the surgical site. He denied any injury to the surgical site. The wound was opened and an estimated 30-mL blood clot was removed. There was no active bleeding and the wound was resutured. There was no evidence of recurrence of the tumor at the 12-month follow-up visit.

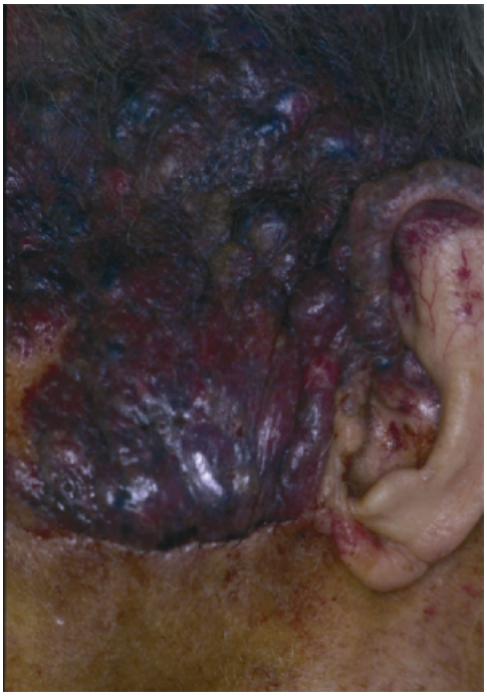

Figure 5. Closure at 1 week postoperatively.

Discussion

The development of basal cell carcinoma in a port-wine stain or nevus flammeus is rare. To the best of our knowledge, 16 cases of these coexisting diseases have been documented. These consist of 14 cases of port-wine stain and 2 cases of nevus flammeus.[5–15] Most of the reported cases, including our case were male patients (13 of 16). The greater incidence of this coexistence in men may be explained by women camouflaging their vascular lesions with cosmetic makeup resulting in decreased sun damage. In 11 of the 16 cases, thorium X, Grenz ray, laser, or carbon dioxide snow were used as the treatment for port-wine stain before the development of basal cell carcinoma.[5,7,9,13–15] There was no previous therapy in the other 5 reported cases.[5,8,10–12] Rogers,[6] Courtemanche,[7] and Shah et al.[13] indicated that the application of thorium X lent a significant contribution to carcinogenesis. Sagi et al.[9] who described a case of basal cell carcinoma arising in port-wine stain following Grenz ray therapy concluded that the possible role of Grenz ray therapy in their case was difficult to determine.

Exposure to sunlight and patients' Fitzpatrick skin type may play a significant role in the development of basal cell carcinoma in both groups with or without prior therapy for their vascular lesions. In our case, as well as these five previously reported cases, basal cell carcinoma developed in untreated port-wine stain. The coexistence of basal cell carcinoma and port-wine stain/nevus flammeus may have occurred by chance.

Considering the treatment of basal cell carcinoma in these patients, the problem of bleeding and hemostasis during and after surgery on port-wine stain is a concern. In this case report, there was no intraoperative problem with hemostasis. Bleeding was controlled with epinephrine in the local anesthesia (0.5% xylocaine with 1:200,000 epinephrine), pressure during surgery, and a pressure bandage postoperatively. The excision of the tumor was carried into the subcutaneous fat on the cheek. No large vessels were identified or bled during surgery. Similarly, there was no excessive bleeding from the port-wine stain in the upper dermis. The wound was closed with subcutaneous interrupted sutures for the close approximation of the dermis. The epidermis was closed with a 6-0 running nylon suture. Despite the adequate closure and the postoperative pressure dressing, the wound bled 6 to 12 hr later and formed a hematoma in the subcutaneous fat. On opening the hematoma, no active bleeding was found. The hematoma was evacuated and the wound resutured without further bleeding or hematoma. Although the source of the hematoma could not be determined with certainty, it is assumed to have been from the port-wine stain. When operating on port-wine stain, the systematic surgical closure and close follow-up especially in the early postoperative period are strongly recommended. Certainly, we advise that surgery of the skin where port-wine stain is involved should be undertaken in a fully equipped facility by physicians experienced in skin surgery.

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