Melanoma In Situ Within a Port-Wine Stain

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PRACTICE **POINTS**

- Nonmelanoma skin cancer is known to develop in port-wine stains, most commonly basal cell carcinoma.
- The range of skin cancer types known to arise in these malformations can be expanded to include melanoma in situ.
- It is important to routinely examine these vascular proliferations for new lesions.

To the Editor:

Port-wine stains (PWSs) are the most common type of vascular malformations. Patients rarely develop cancers in the overlying skin. However, we describe a case of melanoma in situ occurring within a long-standing facial PWS.

A 60-year-old white man with a history of a large unilateral facial PWS covering the right ear, lateral cheek, jaw, and neck presented to clinic with a new dark lesion on the right ear that had been growing for a few weeks or more. His PWS had been previously treated intermittently with a pulsed dye laser (PDL) for decades with variable improvement. He had not undergone any laser procedures in the last 8 months but wanted to restart treatment with the PDL. Upon further discussion, he reported a new darker area on the right earlobe that was growing. He had no personal or family history of skin cancer and was otherwise healthy. Physical examination revealed a large red vascular patch encompassing the ear, cheek, chin, and lateral neck. Within the PWS there was a black and dark brown patch with irregular borders on the right earlobe (Figure 1A). A shave biopsy was performed for histopathologic examination. The biopsy showed a confluent proliferation of atypical melanocytes along the dermoepidermal junction extending down adnexal structures (Figure 2A) that stained positive for MART-1/Melan-A (Figure 2B). In the dermis, solar elastosis and prominent dilated and thin-walled vessels were present. These findings were consistent with a melanoma in situ, lentigo maligna type, overlying a capillary malformation.

The patient underwent a wedge excision of the lesion with 5-mm margins, resulting in a final postoperative size of 2.5×3.5 cm. There was no excessive bleeding with surgery. A delayed repair was done after clear margins were confirmed by pathology (Figure 1B).

Port-wine stains are congenital vascular malformations that affect approximately 0.3% of individuals.[1] Most are located on the head and neck along the distribution of the trigeminal nerve. Cases are thought to occur sporadically, with recent evidence for somatic *GNAQ* mutations in both nonsyndromic cases and in Sturge-Weber syndrome.[2] These lesions become progressively larger with time due to dilation of the capillary proliferation.[3] Melanoma in situ, lentigo maligna type, usually affects white men in the sixth and seventh decades of life. It commonly arises on skin with chronic sun damage, particularly on the head and neck.[4]

Although uncommon, skin cancers have been known to arise in PWSs. Reports of basal cell carcinomas (BCCs) and squamous cell carcinomas (SCCs) have been published, but to date, there are no reports of melanoma or melanoma in situ arising in a PWS. According to a PubMed search of articles indexed for MEDLINE using the terms *melanoma and port wine stain*, *squamous cell carcinoma and port wine stain*, and *basal cell carcinoma and port wine stain*, fewer than 30 cases of BCCs in a PWS and only 4 cases of SCCs in a PWS have been documented, with 1 patient developing multiple BCCs and SCCs.[1,5] Most BCCs (approximately 75%) and SCCs have been associated with historical treatments used to treat PWS before the development of laser therapy, such as grenz

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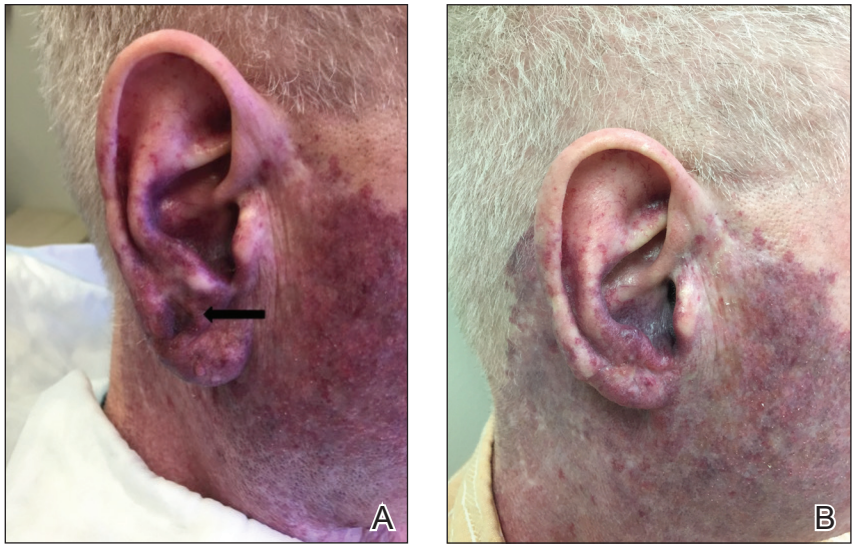

FIGURE 1. Melanoma in situ in a port-wine stain. A, An irregular black and dark brown patch (arrow) on the patient's right earlobe before treatment. B, A good cosmetic outcome was achieved 1 month after wedge excision and repair.

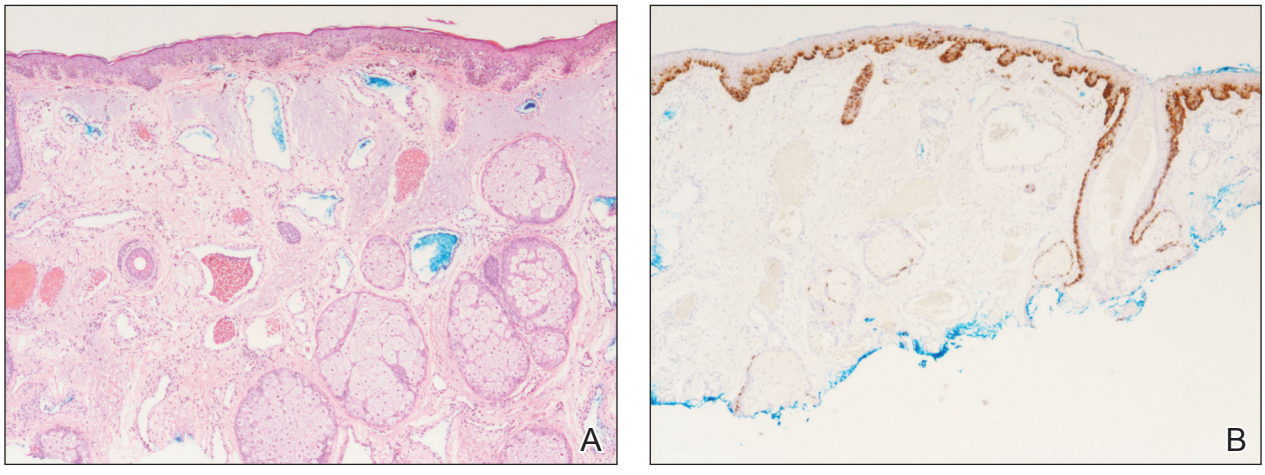

FIGURE 2. Histopathology of a biopsy specimen from the right earlobe. A, A confluent proliferation of atypical melanocytes along the dermoepidermal junction overlying solar elastosis and dilated thin-walled vessels (H&E, original magnification ×100). B, Special staining highlighting the lentiginous spread of atypical melanocytes extending down adnexal structures (MART-1/Melan-A, original magnification ×100).

rays, topical thorium X, and other radiotherapy techniques.[5,6] Interestingly, our patient's PWS had only been treated with a PDL. Other risk factors for skin cancer in a PWS include sun exposure and smoking.[5] There is no evidence that a PDL contributes to the development of skin cancer, but radiotherapy is a major factor.[7]

Treatment of these skin cancers is no different, with both Mohs micrographic surgery and standard excision used when appropriate. Despite the vascular nature of the lesion, there is only a minimal increase in bleeding risk.[3] Most reports indicate no increase in perioperative bleeding.[5,7] One case documented a hematoma developing postoperatively.[6]

This case of melanoma in situ arising in a PWS expands the range of skin cancer types known to arise in these malformations. Because of the potential for skin cancer to develop in a PWS, it is important to routinely examine these vascular proliferations.

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