Accelerated development of conjunctival melanoma during pregnancy may be simply coincidence. This has been postulated for cutaneous melanoma, although most studies suggest a genuine increased risk. The two main postulated theories for this increased risk are hormonal factors and decreased immunity. Several investigators have examined the role of sex hormone receptor status in conjunctival melanoma, PAM, and nevi. Chowers et al. investigated 2 cases of conjunctival melanoma and 13 cases of PAM and were unable to detect estrogen receptors in any. Paridaens et al. found that 6 of 15 conjunctival melanomas were estrogen receptor positive. More recently, Pache et al. found expression of progesterone receptors in 96% of cases, consisting of 69 nevi, 5 cases of PAM, and 2 melanomas. Staining for estrogen and progesterone receptors was negative in this patient.

Changes in immunity that accompany pregnancy may also play an important role. To protect the fetus from rejection, pregnancy induces immunosuppression with a predominant T-helper 2 response to inciting stimuli. Indeed, the severity of many diseases has been shown to be affected by pregnancy. This anti-inflammatory response may also affect tumor surveillance and promote tumor growth. This reduction in immune surveillance has been suggested by Paridaens et al., who described a young woman with growth of conjunctival melanosis and melanoma during the course of 3 pregnancies.

An additional feature of concern in this patient was that the PAM was nonpigmented in some areas. While PAM sine pigmento is well recognized, often in conjunction with pigmented areas, it can be difficult to follow up.

In conclusion, while this link with pregnancy and conjunctival melanoma progression cannot be fully explained, it would be prudent for young women with PAM with atypia to be closely monitored during reproductive life.

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Caruncular Alveolar Rhabdomyosarcoma in a Woman Previously Treated for Breast Cancer

Rhabdomyosarcoma is a malignant mesenchymal tumor that commonly appears in childhood. It is much less frequently seen in adults, and orbital manifestation represents less than 1% of adult cases. We report the case of an alveolar rhabdomyosarcoma that manifested as a caruncular mass in a 54-year-old woman 4 years after treatment for breast carcinoma.

Report of a Case. A 54-year-old woman had a 1-week history of a rapidly growing painless right caruncular mass that did not cause any functional impairment (Figure 1A). Her medical history was significant for a grade II infiltrating ductal adenocarcinoma of the breast 4 years prior. She underwent left mastectomy with axillary lymph node examination, and the results were negative for metastases. She was treated with adjuvant chemotherapy including anthracycline and Herceptin. Following this, she received long-term anastrozole therapy.

The caruncular mass was initially thought to be a metastasis from her breast carcinoma. Magnetic resonance imaging showed an enhancing mass in the medial aspect of the right orbit inferior to the medial rectus, measuring 9 × 11 × 13 mm (Figure 1B). An incisional biopsy was performed, and histopathological analysis showed sheets of medium-sized malignant cells with minimal cytoplasm, pleomorphic nuclei, and numerous mitoses and apoptotic cells, initially thought to be consistent with metastatic ductal adenocarcinoma of the breast. However, immunohistochemical stains were negative for keratin, estrogen and progesterone receptors, and lymphoid and melanocytic markers. The tumor cells stained strongly for vimentin, smooth muscle actin, desmin, myogenin, and MyoD1 (Figure 2).

Although the initial histological analysis suggested embryonal rhabdomyosarcoma, the final diagnosis of alveolar rhabdomyosarcoma was confirmed by fluorescence in situ hybridization analysis; this demonstrated a PAX3-FKHR gene fusion, molecular evidence for the t(2;13) chromosomal translocation that is diagnostic for the alveolar subtype. The prognosis of orbital alveolar rhabdomyosarcoma is worse than that of the embryonal subtype. A complete workup including computed tomography, magnetic resonance imaging, bone scan, and bone marrow biopsy did not reveal any other lesions or metastases.

Despite treatment with vincristine sulfate, cyclophosphamide, and dactinomycin alternating with ifosfamide, mesna, and etoposide every 3 weeks for a minimum of 11 cycles, she had a recurrence and required exenteration 9 months after her initial visit. Although there

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was complete resection with exenteration, she had a recurrence within the right orbit and subsequently metastases to the abdomen, right parotid gland, and left orbit.

Comment. Rhabdomyosarcoma is most commonly seen in the pediatric population, where it is the most common soft-tissue malignant neoplasm of childhood and the most common primary orbital malignant neoplasm of children. Manifestation of primary rhabdomyosarcoma in adults is much less common. A review of primary orbital rhabdomyosarcoma in adults revealed only 14 published cases in the literature, predominantly of the embryonal and pleomorphic types. Our review of the literature did not reveal any case that manifested as a caruncular lesion, although rhabdomyosarcoma of the orbit and sinuses has been reported in an adult patient previously treated for breast cancer.

Uterine rhabdomyosarcomas have been reported in patients treated with either tamoxifen citrate or anastrozole for breast carcinoma. Although a causative effect cannot be elicited, it is interesting that such a rare tumor as rhabdomyosarcoma in adults has been associated with patients previously treated with anastrozole or tamoxifen for breast cancer. However, the role of these medications in the development of rhabdomyosarcoma is purely speculative and requires further investigation before a pathogenic effect can be proposed.

Figure 2. Histological analysis shows a sheet of malignant medium-sized cells (hematoxylin-eosin, original magnification ×100) (A) with minimal cytoplasm, pleomorphic nuclei, and numerous mitoses and apoptotic cells (hematoxylin-eosin, original magnification ×400) (B). Immunohistochemical staining is strongly reactive for desmin (immunoperoxidase with hematoxylin counterstain, original magnification ×400) (C) and MyoD1 (immunoperoxidase with hematoxylin counterstain, original magnification ×400) (D), which are both characteristic of rhabdomyosarcoma.

Figure 1. Photograph demonstrating a right caruncular mass (A), and coronal magnetic resonance image demonstrating the mass (arrow) adjacent to the right medial rectus in the anterior orbit (B).


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