cent scleral channels. The several years between the silicone oil placement and onset of discomfort may be explained by long-term, slow leakage of silicone oil through the sclera.

Free silicone oil within the extracellular matrix commonly causes a chronic granulomatous reaction that may lead to inflammation or functional impairment. Our patient described late development of ocular and orbital pain in a blind eye treated with silicone oil. Enucleation and orbital granuloma removal resolved the pain. It is important to consider extrascleral silicone oil granulomas in patients with ocular and orbital pain following treatment with silicone oil tamponade.

Author Affiliations: Departments of Ophthalmology and Visual Sciences (Drs Couch and Harocopos) and Pathology and Immunology (Dr Harocopos), Washington University, and Departments of Ophthalmology (Drs Couch and Hold) and Otolaryngology–Head and Neck Surgery (Dr Holds), Saint Louis University, St Louis, Missouri.

Correspondence: Dr Couch, 12990 Manchester Rd, Ste 103, Des Peres, MO 63131 (smcouchmd@gmail.com).

Financial Disclosure: None reported.

Funding/Support: This work was supported by Heed Ophthalmic Research Foundation and Research to Prevent Blindness.


Progression of Primary Acquired Melanosis With Atypia During Pregnancy

Primary acquired melanosis (PAM) of the conjunctiva manifests as unilateral patchy areas of pigmentation usually in middle-aged or elderly patients. It can be differentiated histologically by the degree of atypia of melanocytes. Without atypia, PAM is a benign melanocytic proliferation. With atypia, PAM may progress to malignant melanoma. With severe atypia, PAM progresses to melanoma in about 13% of cases. We describe a case of PAM that progressed during pregnancy in a young patient.

Report of a Case. A 28-year-old woman with brown irides had an 8 × 7-mm pigmented area on the right temporal bulbar conjunctiva (Figure 1A). She had noted this several years earlier but felt it was increasing in size. The area was biopsied and showed PAM with moderate atypia (Figure 2A). She was treated with a standard 9-week course of topical mitomycin C, 0.04%, 4 times/d, prednisolone acetate eyedrops, 0.5%, 4 times/d, and ocular lubricants. The pigmented area regressed and remained stable during the next 18 months (Figure 1B). Seven months later, an increase in size and pigment density was noted (Figure 1C). A repeated biopsy showed PAM with moderate atypia (Figure 2B). It was treated with a second course of mitomycin C with good response (Figure 1D).

Figure 1. Clinical photographs. A, Pigmented area of the right temporal bulbar conjunctiva prior to the first course of treatment. Arrow indicates biopsy site shown in Figure 2A. B, The pigmented area regressed following incisional biopsy and after completion of the first course of mitomycin C. C, Increase in the size of the pigmented area before the second course of mitomycin C. Arrow indicates biopsy site shown in Figure 2B. D, The pigmented area again regressed after incisional biopsy and completion of the second course of mitomycin C. E, The lesion showed a definite increase in size when the patient was followed up at 14 weeks’ gestation. Arrow indicates biopsy site shown in Figure 2C. F, Significant improvement in the wound site and in the appearance of the pigmented area 1 month after childbirth, following excisional biopsy and cryotherapy. Arrow indicates biopsy site shown in Figure 2E.
At follow-up 14 months later, definite growth of the area was documented (Figure 1E). The patient was 14 weeks' pregnant. Excision biopsy and cryotherapy were performed. Histologic analysis showed conjunctival melanoma in situ with suspicion of stromal invasion (Figure 2C and D). The lesion was closely monitored for the remainder of the pregnancy.

Seven months later, 1 month after childbirth, there was significant improvement at the wound site (Figure 1F). However, a repeated biopsy performed from a nonpigmented area showed PAM with atypia (Figure 2E and F).

Comment. Changes in cutaneous melanocytic lesions and progression to malignant melanoma during pregnancy are well documented. Similarly, conjunctival nevi have been documented to change during pregnancy. Progression of PAM with atypia during pregnancy has rarely been documented.
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 2 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 pregnancy.

**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 $9 \times 11 \times 13$ 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 (1;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...