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Extensive Morphea-Form Basal Cell Carcinoma Invasion of the Iris and the Orbital Contents*

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The most common malignancy encountered clinically in basal cell carcinoma of the skin involves the eyelid. Usually the lesion is nodular, well localized, circumscribed, and does not metastasize, thus lending itself to relatively simple local excision (1-4). The morphea-like or sclerosing type of basal cell carcinoma, however, features basaloid cells that grow in an Indian-file pattern of cords of cells in a thick, dense, fibrous tissue stroma, like a scirrhous carcinoma of the breast. This growth pattern makes it difficult to determine clinically the extent of the lesion, because the cut end of the biopsied lesion may show only fibrous tissue while tumor cells may lie undetected in the deeper tissue. The morphea-form basal cell carcinoma is thus much more aggressive and invasive than the common “nodular” type. The sclerosing type of basal cell carcinoma may invade the deeper tissue of the eyelid, extend into the orbit, and even into the globe itself.

The following case report is a tragic example of morphea-form basal cell carcinoma that eventually required exenteration of the orbit to effect a “cure.” Invasion of tumor cells into the iris and ciliary body is extremely rare and confirms the clinical impression that morphea-form basal cell carcinoma must be treated vigorously and decisively, preferably by chemosurgery (Mohs’ technique) if available.

Case Report

Clinical history
An 80-year-old woman was admitted to William Beaumont Hospital on April 21, 1985, after she attempted suicide by slashing her wrists. She had been hospitalized in 1981 for a previous suicide attempt via a drug overdose. In 1980 the patient had had surgery for excision of a basal cell carcinoma of the right side of her face. Multiple areas of carcinoma resection left her unable to open the right side of her mouth, chew properly, or brush her teeth. The right eyelids were contracted, and the right cornea was covered shut. An ulcerated skin lesion of the right lower lid looked suspicious for basal cell carcinoma recurrence. A computed tomography scan showed a mass in the right orbit, presumably a tumor (Fig 1).

On April 23, 1985, the patient underwent surgery for multiple facial biopsies; the ulcerated skin lesion of the right cheek was positive for basal cell carcinoma. The patient and her family were advised that clinically it seemed likely that the cancer had invaded the entire right orbit, lower forehead, and glabellar region of the nose, and that if treatment were to be definitive, a wide resection of the right side of the face and exenteration of the right orbit might be required later.

On April 30, 1985, lesions of the right cheek, the right upper lip, the upper and lower right eyelids, a portion of the right forehead, nose, and left medial canthus were excised. The excision process was guided by microscopic frozen section biopsies. With the evidence of orbital invasion, proven by biopsy, arrangement was made with two ophthalmic plastic surgeons to exenterate the right orbit. On May 6, 1985, the right orbit was exenterated and biopsies were taken from the bony floor of the orbit. Although the bony orbit was found microscopically to be free of basal cell carcinoma, the entire globe and retrobulbar tissues were infiltrated with the cancer. The excised areas received skin grafts, and the cosmetic result was excellent at three-months follow-up.

Pathology
The globe was encased in a conical segment of soft tissue, which in areas was firm and of a gray-white color. The globe itself measured 22 mm horizontally, 23 mm vertically, and 27 mm anteroposteriorly.

At the limbus, basal cell carcinoma had invaded the cornea and sclera along the emissary vessels and had reached the anterior chamber angle, the ciliary body, and the peripheral iris. Free basal cell carcinoma cells were floating in the anterior chamber aqueous, as though in a tissue culture medium. The iris stroma was diffusely invaded by basal cell carcinoma, and squamous differentiation (baso-squamous cell carcinoma) within nests of basal cells was noted on the anterior iris and the base of...
Eig 2—Invasion of sclera, ciliary body, and iris by carcinoma (hematoxylin-eosin stain) (X40).

The ciliary body (Figs 2 through 4). Hemorrhage occurred in the ciliary body and the iris. The basal cell carcinoma had invaded the extraocular muscles, Tenon’s fascia, and the sclera in the posterior portion of the globe.

Basal cell carcinoma cells also had invaded the optic nerve sheaths posterior to the globe. Strands and cords of basalloid cells had invaded the orbital fascia and fat, like scirrhous carcinoma of the breast, and extended to the cut margin of the specimen (Fig 5).

Diagnosis

The following diagnoses were made: 1) morphea-form basal cell carcinoma with invasion of the limbus, anterior chamber angle, iris, and ciliary body; and 2) morphea (fibrosing type) basal cell carcinoma with invasion of the posterior sclera, extraocular muscles, Tenon’s fascia, and optic nerve sheath.

Fig 4—Squamous differentiation within basal cell carcinoma on anterior iris (hematoxylin-eosin stain) (X200).

Fig 5—Basaloid cells within sclera and orbital connective tissue at distal end of exenteration specimen indicating incomplete excision (hematoxylin-eosin stain) (X200).

Fig 3—Basal cell carcinoma tumor on anterior iris with malignant cells in the anterior chamber (hematoxylin-eosin stain) (X100).
Discussion

When a lesion is diagnosed as a morphea-form type of basal cell carcinoma, proper treatment involves surgical removal of the lesion with complete microscopic control. The proper technique involves excising the tumor in successive layers and submitting the serial sections to the pathologist in the operating room for examination by frozen sections. By using total microscopic control, the Indian-file pattern of basal cell carcinoma can be followed to its proper termination, thus eliminating the possibility of missing cancerous tissue invisible to the naked eye and not found by palpation.

The "fresh-tissue" technique for frozen section and serial excision of the tumor can be supplemented by the fixed-tissue technique in which the tissue is chemically fixed in situ (eg, zinc chloride-type fixative) before successive layers of the tissue are excised.

The Mohs' procedure of microscopic surgery for the microscopically controlled excision of eyelid carcinoma is probably the best technique currently available. Failure to recognize the exact extent of morphea-form basal cell carcinoma is common, and special attention must be given to treating such lesions properly.

Summary

Although basal cell carcinoma is the most frequent malignancy encountered in the adult eyelid, we report a rare case of basal cell carcinoma invasion of the iris. The lesions are usually well localized and nodular, but the fibrosing type (morphea-form) of basal cell carcinoma is clinically deceptive in its growth pattern. The morphea form of basal cell carcinoma tends to invade the dermis and the orbit like scirrhous carcinoma of the breast, which makes the extent of the lesion difficult to determine. An aggressive surgical approach (eg, the Mohs' procedure) is required to forestall recurrence of the tumor (5-13).

References