Inadvertent Evisceration of Eyes Containing Uveal Melanoma

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Objectives: To report an important complication of ocular evisceration therapy for blind, painful eyes that has been unreported in the literature, and to stress the need for careful preoperative evaluation to exclude occult neoplasms prior to therapy.

Design: Multicenter, retrospective, nonrandomized clinicopathological case series of patients found to have previously unsuspected uveal malignant melanoma during histopathological examination of blind, painful eyes treated by evisceration.

Results: Histopathological examination of evisceration specimens disclosed previously unsuspected uveal melanoma in 7 patients who were treated for blind, painful eyes. Inflammation caused by necrosis of the tumor and other ocular tissues led to misdiagnosis as endophthalmitis, orbital cellulitis, or idiopathic orbital inflammation in several cases. Preoperative imaging was not performed in 3 cases and failed to detect tumors in the remaining 4 cases. Failure of necrotic tumors to enhance contributed to misdiagnosis.

Conclusions: The presence of a malignant intraocular neoplasm should be excluded prior to evisceration of any blind eye or blind, painful eye, particularly with opaque media. Necrosis-related inflammation can confound the clinical diagnosis of occult lesions, as can failure of necrotic tumors to enhance on imaging studies.


THERE IS RENEWED INTEREST in the evisceration of blind, painful eyes among American oculoplastic surgeons. This has been sparked by the procedure’s purported superior cosmesis and articles downplaying the risk of sympathetic uveitis.\(^1\)\(^2\) Based on our collective experience, we believe that the risk of inadvertently eviscerating an eye containing an unsuspected malignant neoplasm probably is greater than the risk of sympathetic uveitis. This complication appears to be underrepresented in the literature as only 2 cases of melanoma diagnosed after evisceration have been reported since 1967.\(^3\)\(^4\) Recent trends in ophthalmic practice have the potential to place more patients at risk for this complication. Herein we report 7 additional cases of uveal melanoma diagnosed in evisceration specimens. Several of these patients were evaluated and treated at university-affiliated eye hospitals where they received preoperative imaging studies. Two patients had orbital inflammatory signs related to tumor necrosis and were treated for inflammatory pseudotumor and/or orbital cellulitis prior to evisceration.

REPORT OF CASES

CASE 1

A chronically ill 89-year-old white woman had progressive visual loss in her left eye of 3 months’ duration. Evaluation by a retinal specialist revealed no light perception OS and an intraocular hemorrhage. Examination at a university eye hospital revealed an intraocular pressure of 70 mm Hg OS, swelling and erythema of her left eyelids, and severe conjunctival injection and chemosis (Figure, A). There was no view of the left fundus. The right eye was normal. She was thought to have neovascular glaucoma secondary to ocular ischemic syndrome. In addition, the...
severity of the adnexal inflammation raised concern about orbital cellulitis or an idiopathic inflammatory pseudotumor. An oculoplastic surgeon admitted her emergently for antibiotic therapy prior to evisceration.

Computed tomography of the left eye and orbit was obtained on admission (Figure, B). Radiological diagnoses included cellulitis vs dacyroadenitis of the left orbit, a hyperdense left vitreous, and choroidal thickening representing a mass or detachment. The paranasal sinuses were uninvolved and there was no evidence of an orbital abscess. The posterior choroidal thickening was disregarded because there was no abnormal enhancement.

The patient’s eye was eviscerated after an empirical preoperative course of intravenous antibiotics and methylprednisolone sodium succinate. Histopathological examination disclosed an extensively necrotic melanoma of mixed cell type (Figure, C and D). The iris and ciliary body were necrotic and there was florid iris neovascularization.

The scleral shell and implant were later removed by an ocular oncologist who biopsied 4 quadrants of orbital tissue. Histopathological examination disclosed no residual tumor in the scleral shell or orbital biopsies. Six days postoperatively, the patient died suddenly of a presumed thromboembolic event. A postmortem examination was not performed.

CASE 2

A 70-year-old white man visited a university eye hospital with severe pain in his left eye. Twenty years previously he had retinal reattachment surgery after ocular trauma. The pain began 8 months previously and had increased in severity 2 months prior to the initial visit. Examination revealed a scarred, disorganized left globe with no light perception. He was diagnosed with phthisis bulbi secondary to trauma and surgery, and his eye was eviscerated by an oculoplastic surgeon. Preoperative imaging studies were not performed.
Histopathological examination revealed an extensively necrotic uveal melanoma of mixed cell type. The scleral shell subsequently was removed and histopathological examination disclosed focal tumor infiltration of the optic nerve and subarachnoid space. The patient died suddenly of a cerebrovascular accident 3 months postoperatively. The latter was thought to be unrelated to his ocular tumor, although a postmortem examination was not performed.

CASE 3

A 70-year-old-Hispanic woman with a blind, painful eye attributed to glaucoma and vitreous hemorrhage was referred to an oculoplastic surgeon. Her eye was eviscerated the next day without having preoperative ultrasonography or imaging studies performed. Histopathological examination disclosed confluent sheets of spindle and epithelioid malignant melanoma cells that were immunoreactive for melanoma markers S-100 protein and HMB-45. Occasional mitotic figures were encountered. Histopathological examination of the scleral shell disclosed no residual melanoma. No follow-up data are available.

CASE 4

A chronically ill 87-year-old white man with severe visual loss in his right eye and urine cultures positive for methicillin-resistant Staphylococcus aureus was referred to an oculoplastic surgeon at a university eye hospital with the diagnosis of orbital cellulitis. Examination showed right ocular proptosis, periorbital edema, ocular injection, and a right afferent papillary defect (Figure, E). Computed tomography disclosed a largely intraconal, 27-mm-diameter orbital mass abutting the posterior surface of the globe (Figure, F). Although the patient was afebrile with a white blood cell count of 7100/µL, he was diagnosed as having endophthalmitis, orbital cellulitis, and an orbital tumor. After intravenous antibiotic therapy, the right globe was eviscerated and the orbital tumor was biopsied. Histopathological examination disclosed an extensively necrotic mixed cell—type uveal melanoma (Figure, G and H). Extensive fibrosis, hemosiderin pigment, and osseous metaplasia were indicative of chronicity. The “orbital tumor” was a massive focus of extracocular extension from the primary uveal melanoma. The patient’s inflammatory signs were attributed to extensive necrosis of the tumor and intraocular structures caused by glaucoma. Follow-up data are not available.

CASE 5

A 67-year-old man had a history of ocular melanoma in his left eye treated with radiation therapy 15 years previously. He developed a corneal abscess and the left eye was eviscerated. Preoperative imaging was not performed. It is not known whether the surgeon was aware of the patient’s history of melanoma. Microscopical examination disclosed fragments of mixed cell—type uveal melanoma. The tumor cells appeared viable but no mitotic figures were observed. The cornea and vitreous were acutely inflamed, and blood, blood-breakdown products, cholesterol, and collagenized fibrovascular tissue were associated with the tumor. No follow-up data are available.

CASE 6

A 70-year-old-Hispanic woman with a blind, painful eye attributed to glaucoma and vitreous hemorrhage was referred to an oculoplastic surgeon. Her eye was eviscerated the next day without having preoperative ultrasonography or imaging studies performed. Histopathological examination disclosed confluent sheets of spindle and epithelioid malignant melanoma cells that were immunoreactive for melanoma markers S-100 protein and HMB-45. Occasional mitotic figures were encountered. Histopathological examination of the scleral shell disclosed no residual melanoma. No follow-up data are available.

COMMENT

Evisceration has become an increasingly common treatment option for patients with blind, painful eyes. Advantages of evisceration over enucleation include ease of surgery, the barrier effect of the intact sclera in preventing orbital spread of infection in cases of endophthalmitis, and perceived functional and cosmetic benefits. In a review of 1028 eyes enucleated or eviscerated during a 20-year period, Hansen et al5 noted a significant increase in the number of eviscerations performed in recent years, which they attributed to the perceived cosmetic advantage of this procedure.

Renewed interest in evisceration was sparked by an article published in 1999 by Levine and coworkers that concluded that “evisceration is an effective and safe procedure with a low risk for sympathetic uveitis.”11 In the past, a commonly expressed objection to evisceration was the increased risk of sympathetic ophthalmia in traumatized eyes that were eviscerated rather than enucleated. Green et al9 reported 4 cases of sympathetic ophthalmia occurring after evisceration of the fellow eye, and 6 additional cases were reported in Japan.7 Evisceration theoretically provides less adequate prophylaxis against the development of sympathetic ophthalmia because the pro-
procedure leaves antigenic material behind in intrascleral emissarial canals.

Sympathetic ophthalmia following evisceration does appear to be a real entity, but fortunately it occurs very rarely. None of us have personally seen a case. Based on our collective experience reported herein, we believe that the risk of accidentally eviscerating an eye containing an unsuspected intraocular tumor probably is greater than the risk of sympathetic ophthalmia. We are aware of several other cases of melanoma that were eviscerated elsewhere as well as a number of unsuspected nonmelanocytic neoplasms including choroidal lymphoma, a spindle cell neoplasm, carcinoma of the nonpigmented ciliary epithelium, an anaplastic tumor, and 2 cases of retinoblastoma. One of the latter, a 4-year-old girl erroneously thought to have Coats disease, was described by David J. Apple, MD, at the 1992 meeting of the Verhoeff Zimmerman Society.8

We believe that this complication has been grossly underrepresented in the literature due to medicolegal concerns. Starr and Zimmerman9 described 2 cases of uveal melanoma that were treated by evisceration in 1962. Both patients were incorrectly diagnosed as having panophthalmitis and both died of recurrent orbital melanoma, 2 and 4.3 years after evisceration. Only 2 other cases of melanoma diagnosed after evisceration have been reported since 1967.3,4

Before the availability of modern imaging techniques such as ultrasonography, computed tomography, and magnetic resonance imaging, 10% of blind, painful eyes with opaque media were found to contain unsuspected malignant neoplasms, usually uveal melanomas, on pathological examination.10,11 Although the number of unsuspected tumors has markedly decreased in recent years, ophthalmic pathologists still discover clinically unsuspected neoplasms during the pathological examination of blind, painful eyes.

We believe that recent trends in ophthalmic practice have the potential to place more patients at risk for this complication. Comprehensive ophthalmologists are removing fewer eyes. Instead, they are referring patients to oculoplastic surgeons who often perform the surgery after little or no clinical workup with no more knowledge of the case than that the patient has a blind, painful eye. The consulting surgeon also is the person making the decision as to whether an enucleation or evisceration is truly responsible.

It is imperative to perform an adequate preoperative evaluation prior to enucleation. This should include obtaining a good history and imaging, preferably B-scan ultrasonography, if there are opaque media. If melanoma is found in an enucleation specimen, the scleral shell should be removed and the surrounding orbital tissue should be biopsied. If the orbital biopsies are positive, orbital exenteration or radiation therapy are indicated.

Our study shows that preoperative imaging does not totally exclude the possibility of inadvertently eviscerating an eye with occult melanoma. Forty-five years ago, Frederick C. Blodi, MD, made the following comment about the risk of sympathetic ophthalmia after evisceration: ‘Whether we want to take the risk of this rare—

contrast enhancement also has its proponents. However, many believe that B-scan ultrasonography performed by a skilled ultrasonographer is the superior technique to exclude an intraocular tumor in a blind eye with opaque media. If a mass is found on ultrasonography, which may be the case when there are disorganized intraocular contents, the study should be reviewed by an experienced ocular oncologist if evisceration is considered. This can be done by e-mailing the images. If the oncologist cannot exclude the possibility of an intraocular tumor, the eye should be enucleated.

Imaging studies must be interpreted carefully in patients who have inflammatory signs that might be caused by necrotic tumors. Two of our cases demonstrate that orbital inflammation may be the initial manifestation of necrotic uveal melanoma. In such patients, the orbital inflammation probably is caused by infarction of the tumor and other intraocular structures in an eye with severe neovascular glaucoma. Although inflammation is an unusual initial manifestation of uveal melanoma, necrotic choroidal melanomas clearly have been shown to cause orbital inflammation.12,35 Rose et al36 described 3 patients with intraocular malignant neoplasms with initial manifestation as orbital inflammation. Lea et al37 also reported a necrotic intraocular tumor manifesting as cellulitis and emphasized that computed tomographic scanning can be misleading in the diagnosis of a tumor after spontaneous necrosis.16 The 2 cases described by Starr and Zimmerman9 were both initially misdiagnosed as panophthalmitis. The first case in our series was given the presumptive diagnoses of orbital cellulitis vs inflammatory pseudotumor. Improvement on systemic antibiotic and corticosteroid therapy and failure of the necrotic tumor to enhance on computed tomography were factors that contributed to misdiagnosis.

The effect of inadvertent evisceration of uveal melanoma on the patient's prognosis is not specifically known, although it is likely detrimental. The physician who eviscerates unsuspected uveal melanoma is at risk as well. He or she could be found liable for failure to diagnose and failure to offer the patient effective treatment options if the patient subsequently develops metastases. The latter risk certainly is not inconsequential; overall, a patient with conventionally treated melanoma has approximately a 50% lifetime risk of dying from metastatic melanoma. Therefore, the eviscerating physician has 1 chance in 2 of being blamed, regardless of whether he or she is truly responsible.

It is imperative to perform an adequate preoperative evaluation prior to evisceration. This should include obtaining a good history and imaging, preferably B-scan ultrasonography, if there are opaque media. If melanoma is found in an evisceration specimen, the scleral shell should be removed and the surrounding orbital tissue should be biopsied. If the orbital biopsies are positive, orbital exenteration or radiation therapy are indicated.
but certainly occurring—severe complication, against an alleged cosmetic advantage, is up to the conscience of every ophthalmologist.”

Dr Blodi’s comment is equally applicable to the risk of an occult tumor. Enucleation is the safest treatment for a blind, painful eye.

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REFERENCES


Ophthalmological Ephemera

In 1795, Dr Isaac Thompson concocted an eye water of zinc sulfate, saffron, camphor, and rose water. It was sold as late as 1939. This is 1 of a series of 32 medical trade cards advertising the product from 1875 through 1895.

Courtesy of: Daniel M. Albert, MD, MS.