Adenoid Cystic Carcinoma of the Lacrimal Gland Simulating a Dermoid Cyst in a 9-Year-Old Boy

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Adenoid cystic carcinoma of the lacrimal gland is a malignant neoplasm that is generally found in adults and is usually managed by orbital exenteration and supplemental external beam irradiation or chemotherapy. A recent report has suggested that the tumor may have a less malignant course in children. We describe a case of adenoid cystic carcinoma of the lacrimal gland that simulated a dermoid cyst clinically and radiographically in a 9-year-old boy. The patient was treated with local surgical resection of the mass, followed by orbital plaque brachytherapy. Based on a review of the literature and our recent experience, the advisability of a more conservative approach to this tumor in selected cases is discussed. Although no prognostic conclusions can be drawn on the basis of a single case report with short follow-up, the relatively earlier detection of this tumor made possible by modern orbital imaging studies may allow total removal at an earlier stage and prevent orbital exenteration in a patient with normal vision. Recent developments suggest that there may be a basis for reassessing the advisability of a radical approach to the management of adenoid cystic carcinoma of the lacrimal gland in selected cases.


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Adenoid cystic carcinoma is the most common primary malignant tumor of the lacrimal gland, but it accounts for less than 4% of all lacrimal gland lesions.1-3 It is a malignant neoplasm that has generally been associated with a poor prognosis.4-11 Consequently, there has been a tendency for clinicians to recommend radical management, with orbital exenteration, irradiation, or chemotherapy. Authorities have expressed pessimism regarding patient survival despite such radical measures.5-11 Adenoid cystic carcinoma is diagnosed at a mean age of about 40 years5 and rarely in childhood.12,13 It has been suggested that adenoid cystic carcinoma in children may be associated with a better prognosis than previously believed.13

We describe herein a clinicopathologic correlation of an orbital mass that had radiographic features suggestive of a dermoid cyst but that proved histopathologically to be an adenoid cystic carcinoma of the lacrimal gland in a 9-year-old boy. To our knowledge, he is the second youngest patient ever recorded with this tumor. The patient did not undergo orbital exenteration but was treated with local excision of the tumor followed by orbital brachytherapy.14 Although long-term follow-up is lacking, we present an argument, based on recent developments, for reassessing the advisability of orbital exenteration for all cases of adenoid cystic carcinoma of the lacrimal gland, particularly in younger patients.

REPORT OF A CASE

A previously healthy 9-year-old boy was referred to us on April 4, 1997, with a 3-month history of downward displacement of the left eye and mild pain above the left eye. He had undergone heart surgery at age 2 years because of aortic stenosis but his medical history was otherwise unremarkable. Uncorrected visual
acuity was almost 20/20 OU and there was no diplopia or proptosis. He had 1 mm of left blepharoptosis and slight swelling of the subcutaneous tissue superotemporally (Figure 1). A smooth, fluctuant, slightly tender, subcutaneous mass was palpable along the superolateral orbital rim.

Orbital computed tomography (CT) revealed an ovoid, cystic mass in the superotemporal aspect of the orbit anteriorly (Figure 2). It was producing a smooth fossa in the orbital bone superiorly in the area of the zygomaticofrontal suture. The more central cystic portion of the mass showed no contrast enhancement, but the peripheral portion showed some enhancement. Magnetic resonance imaging was not done because of the presence of a metallic wire from prior cardiac surgery. The primary diagnosis based on clinical findings and CT was dermoid cyst. The differential diagnosis also included inflammatory pseudotumor (dacryoadenitis), pleomorphic adenoma, and adenoid cystic carcinoma of the lacrimal gland, but the latter diagnoses were considered unlikely.

Because we have successfully removed a number of benign circumscribed anterior orbit lesions through a conjunctival route, we elected to remove the mass through a superotemporal fornical conjunctival incision. A fairly well-defined mass was removed by soft tissue dissection without complications and the patient had an uneventful postoperative course.

Grossly the specimen was a soft, partly encapsulated mass that measured 21 × 10 × 9 mm. The sectioned specimen showed a partially cystic mass with large spheroidal globules of translucent lipidlike material in the lumen (Figure 3). Microscopically, there were areas of normal lacrimal gland with foci of chronic inflammation. Most of the lesion was composed of round to oval lobules of tumor that showed a cribriform pattern, comprising prominent pools of mucin encompassed by delicate strands of malignant epithelial cells (Figure 4). Some of the tumor lobules appeared to float freely in the cystic portion of the tumor. Cords and nests of tumor cells infiltrated the capsule and reached its outer surface in some sections. There was no appreciable solid basaloid pattern and no convincing evidence of perineural invasion. The final diagnosis was well-differentiated adenoid cystic carcinoma of the lacrimal gland.

The patient was referred for a more extensive pediatric oncology evaluation, which revealed normal findings, with normal chest x-ray and blood chemistry test results. Opinions regarding further management were obtained by telephone consultations with several authorities on orbital diseases and radiation oncology in the United States and Canada. There was a consensus that orbital exenteration should not be done and the parents agreed to additional surgical exploration, biopsy, and orbital brachytherapy.

A superolateral eyelid crease incision was performed with an extra-periosteal approach. After a circular incision through the periosteum, the superotemporal aspect of the orbit was inspected and no suspicious tissue was identified. An en bloc excision of the periosteum and the underlying fat and orbital tissues was performed and marginal areas were submitted for frozen sections. One margin was positive for adenoid cystic carcinoma, but wider excision showed that all margins were negative. Small random biopsy specimens were negative for tumor.

According to a plan designed in conjunction with our radiation oncologists, a 15-mm-diameter round gold plaque without radiation seeds was sutured to the sclera on the superotemporal aspect of the globe to shield the eye from radiation. An custom-designed unshielded radioactive iodine 125 plaque was then placed in the superotemporal orbit to deliver irradiation to the bone and adjacent soft tissues in the superotemporal and posterior aspects of the orbit. The plaque measured 25 × 20 mm and delivered 53 Gy to a depth of 3 mm at a rate of 0.56 Gy/h. The choice of the isotope and the dose distributions were based on a thorough review of the clinical and histopathologic findings by radiation oncologists experienced in ocular brachytherapy. The plaque was subsequently removed surgically. One year after the initial presentation, the patient has a visual acuity of 20/20 OU and no local or systemic recurrence of his tumor. Close long-term follow-up is planned.

COMMENT

Adenoid cystic carcinoma of the lacrimal gland has a tendency to recur...
many years after incomplete excision, exhibit local invasion, and sometimes metastasize. Consequently, there has been a tendency in recent years to employ aggressive treatment with orbital exenteration and removal of adjacent orbital bone (radical orbitectomy) combined with irradiation or chemotherapy.\textsuperscript{1-11} Even with this radical approach, some authorities have expressed concern about the poor prognosis of patients with this tumor.\textsuperscript{5,7} Unfortunately, this tumor can grow slowly and exhibit recurrence and metastasis years after the initial treatment.\textsuperscript{5,8} Therefore, long-term follow-up of treatment methods has been difficult to obtain.

Much of the information that has prompted the pessimism regarding the prognosis for patients with adenoid cystic carcinoma was derived from long-term follow-up of patients treated years ago. Many of the affected patients were seen prior to the development and refinement of modern imaging techniques such as CT and magnetic resonance imaging, which have led to earlier diagnosis while the tumor is still circumscribed. We are aware of several cases in which similar imaging studies, often performed for headache or other unrelated symptoms, have allowed removal of adenoid cystic carcinoma at a much earlier stage than was possible in the past. Consequently, we have received referrals and telephone calls from ophthalmologists who point out that they have excised a circumscribed adenoid cystic carcinoma that seems to have been removed entirely, based on surgical observations and histopathologic studies. According to most reports in the literature,\textsuperscript{9,11} such patients should usually be treated with orbital exenteration and possible irradiation or chemotherapy. However, it seems difficult to justify orbital exenteration in a young person who has excellent vision and no evidence of residual tumor.

The case cited here is an example of the dilemma in the diagnosis and management of adenoid cystic carcinoma of the lacrimal gland. Our patient was 9 years old and had a fluctuant, superotemporal orbital lesion with CT features suggestive of a cyst with bony fossa formation. These features all suggested a diagnosis of orbital dermoid cyst and the diagnosis of adenoid cystic carcinoma was not realized until the pathologic sections were studied. The young age of our patient was also in keeping with a dermoid cyst.

Although this single case report is insufficient to draw conclusions regarding management, we are aware of several similar cases in which the clinician has been reluctant to recommend orbital exenteration when the tumor has apparently been totally removed. Therefore, it is possible that we should reassess our treatment philosophy regarding malignant tumors of the lacrimal gland. Certainly for advanced or recurrent tumors, orbital exenteration is probably appropriate. However, for those cases that are being discovered at an early stage and in which the tumor appears to be removed completely, there may be an argument for avoiding exenteration. This approach may be applicable to all cases in which the tumor appears to have been completely removed surgically, but especially for children with tumors that lack basaloid features or neural invasion on histopathologic examination.\textsuperscript{11,13} Others have propagated a similar conservative philosophy for the management of adenoid cystic carcinoma of the lacrimal gland.\textsuperscript{14}

In the case reported here, the primary diagnosis was dermoid cyst based on the patient’s age, clinical findings, and CT findings. Hence, a conjunctival approach, which is often very effective for removing circumscribed anterior orbital tumors, seemed to be justified. In retrospect, if adenoid cystic carcinoma had been a strong diagnostic consideration, a superolateral orbitotomy with a cutaneous extra-periosteal approach may have been preferable. Although most lacrimal gland lesions are benign, we now believe more firmly that any circumscribed superotemporal orbital tumor should be considered a possible malignant neoplasm of the lacrimal gland until proven otherwise, despite atypical CT findings or young age of the patient.\textsuperscript{12,13}

We have some concerns about the traditional teaching regarding the surgical approaches to lacrimal gland malignant neoplasms. It has been taught that when a malignant epithelial tumor of the lacrimal gland...
is suspected on the basis of clinical findings and imaging studies, a biopsy should be performed through the eyelid and orbital exenteration should be carried out after permanent section confirmation of the diagnosis. We cannot uniformly accept such a rigid dictum. If a suspected lacrimal gland neoplasm is reasonably well circumscribed, a meticulous attempt to remove the tumor completely should be undertaken. The same philosophy should also apply to any orbital mass that appears to be surgically resectable based on imaging studies. We are aware of several cases in which the surgeon felt compelled to do an incisional biopsy to make a diagnosis and then to refer the patient for a second surgical procedure to remove the mass. Because such masses can be malignant, it is probably more prudent to manage any circumscribed mass by excisional biopsy if possible.

In view of the histopathologic findings, orbital brachytherapy was selected as supplemental treatment in this case. Ocular brachytherapy has been used for years to treat selected choroidal melanoma and retinoblastoma. We have used orbital brachytherapy for 2 other orbital neoplasms, but we do not have sufficient follow-up data to comment on its role for such tumors. Subsequent to the treatment of our case, a series of orbital tumors treated with brachytherapy was reported, including 7 cases of adenoid cystic carcinoma. The authors did not provide the ages of the patients with adenoid cystic carcinoma of the lacrimal gland. Although their follow-up is still insufficient, the authors expressed preliminary enthusiasm for the technique. In our case, brachytherapy with a custom-designed radioactive plaque seemed to be an appropriate option to orbital exenteration or simple observation. Intratraicle chemotherapy has also been recently advocated in the supplemental management of this tumor. In our patient, close follow-up is planned, and if there is tumor recurrence, exenteration may be necessary.

Adenoid cystic carcinoma of the lacrimal gland is a rare neoplasm and complete, long-term follow-up on many cases may be difficult to achieve. However, in view of the improved diagnostic and therapeutic methods currently available, a more conservative approach to the management of malignant epithelial tumors of the lacrimal gland should be considered in selected cases.

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REFERENCES