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Adenoid Cystic Carcinoma of the Lacrimal Gland: A Case Report with a Review of the Literature

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ABSTRACT

Adenoid cystic carcinomas, the most common malignancies of the lacrimal gland, are rare overall. We describe a patient who presented with right periorbital swelling developing over 5 months and magnetic resonance imaging findings of a soft tissue mass in the lacrimal fossa with invasion of the adjacent bone. The patient underwent right lateral orbitotomy with tumor debulking. Pathologic analysis showed neoplastic cells in a predominantly cribriform pattern, and the patient was diagnosed with an adenoid cystic carcinoma of the lacrimal gland. We review the clinical, radiographic, and histopathologic features of these rare, aggressive malignancies as well as treatment options with reference to the current literature.

Keywords: adenoid cystic carcinoma; lacrimal; oncology; pathology; tumor

RÉSUMÉ

Le carcinome adénoïde kystique, la plus commune des tumeurs malignes des glandes lacrymales, demeure quand même rare. Nous décrivons le cas d'un patient présentant un œdème périorbital de l'œil droit développé sur une période de cinq mois et les conclusions de l'examen IRM d'une masse de tissu mou dans la fosse lacrymale avec invasion de l'os adjacent. Le patient a subi une orbitotomie latérale droite avec réduction tumorale. L'analyse pathologique a montré la présence de cellules néoplasiques en présentation principalement cribriforme, et le patient a reçu un diagnostic de carcinome adénoïde kystique de la glande lacrymale. Nous examinons les caractéristiques cliniques, radiographique et histopathologiques de ces tumeurs malignes rares et agressives ainsi que les options de traitement, avec des références à la documentation scientifique actuelle.

Introduction

Malignant tumors of the lacrimal gland are rare and have an estimated incidence of 0.073 per 100,000 individuals annually [1]. Adenoid cystic carcinomas are the most common type of lacrimal gland malignancy. Patients with adenoid cystic carcinomas may present with asymmetric facial pain or swelling, diplopia, decreased visual acuity, or ptosis [2, 3]. Magnetic resonance imaging (MRI) is the preferred imaging modality and often shows a nodular, irregular mass that may invade adjacent nerves or bone. Adenoid cystic carcinomas have specific pathologic features that correlate with prognosis. In general, adenoid cystic carcinomas are aggressive tumors with poor prognoses. Treatment most commonly includes surgery with or without radiation and chemotherapy.

We describe a patient who presented with right periorbital swelling and MRI findings of an irregular, soft tissue mass in the lacrimal fossa with globe deformation and bone invasion. The tumor was resected, and pathologic analysis showed an adenoid cystic carcinoma of the lacrimal gland. We review the clinical, radiographic, and pathologic features of these rare malignant tumors as well as treatment options.

Case Presentation

A 39-year-old man with no past medical history presented with swelling around his right eye that had progressively worsened over the preceding 5 months. He had been experiencing

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diffuse headaches for 1 year and double vision for 3 weeks. He denied localized pain or numbness around his eye, weakness in his face, or decreased vision. On examination, the patient had proptosis of the right orbit and nontender edema of his right eyelids and periorbital soft tissue. He had dysconjugate upward gaze. Visual acuity was 20/20 in both eyes, and visual fields were intact. His pupils were equally round and reactive, and the fundoscopic examination was unremarkable. He had no lymphadenopathy. A computed tomography (CT) scan of the face showed a lacrimal mass with bony remodeling. MRI found an enhancing, T1-isointense and T2-hyperintense irregular soft tissue mass in the right lacrimal fossa associated with deformation of the posterolateral globe, downward displacement of the right superior rectus muscle, and invasion of the adjacent orbital walls (Figures 1 and 2).

There was no perineural or intracranial extension. Imaging was performed on a 1.5T GE Genesis Signa scanner using a standard orbital protocol including fat-suppressed T2-weighted and post-contrast T1-weighted images obtained at 3-mm slice thickness in the axial and coronal planes. The patient underwent right lateral orbitotomy and tumor debulking. It was noted intraoperatively that there was no clear demarcation between tumor and normal tissue. Based on the invasion and erosion of nearby bone, the tumor was given a T4 classification.

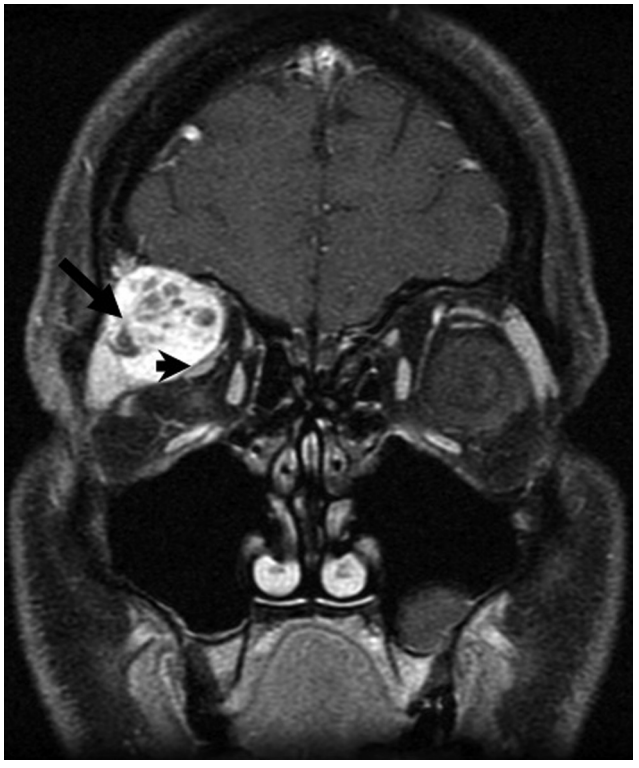

Figure 1. T1-weighted post-gadolinium fat-suppressed MRI showing an enhancing soft tissue mass (arrow) in the right lacrimal fossa with downward displacement of the right superior rectus muscle (arrowhead) and invasion of the adjacent bone.

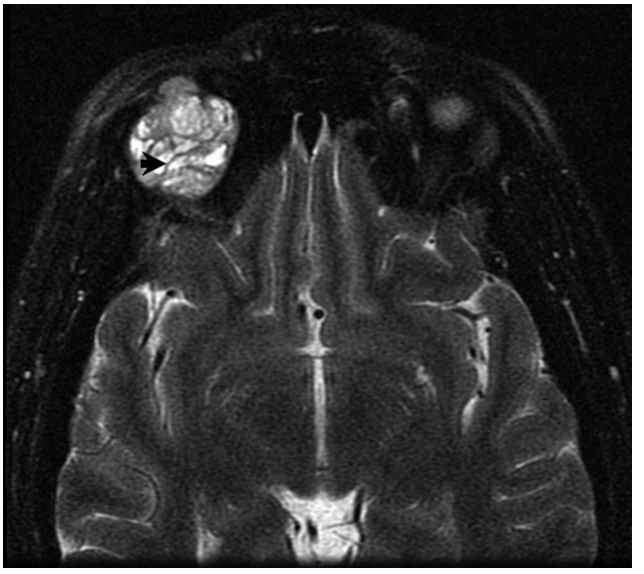

Figure 2. T2-weighted fat-suppressed MRI showing a hyperintense, irregular mass with linear septations (arrowhead).

On histologic analysis, the tumor appeared to infiltrate the adjacent benign lacrimal gland tissue. It contained round, pseudocystic structures filled with basophilic glycosaminoglycans and eosinophilic, hyalinized basal lamina (Figure 3). Neoplastic cells were predominantly in a cribriform pattern, and less than 30% were in solid sheets. Occasional tubular structures were noted. Smooth muscle actin and CD117 staining identified a predominant myoepithelial cell population (Figure 4), confirming the diagnosis of intermediate-grade lacrimal gland adenoid cystic carcinoma. No perineural or lymphovascular invasion was identified.

Postoperatively, the patient was referred for radiation therapy. He was subsequently lost to follow-up. The cribriform growth pattern of his tumor and the absence of neural

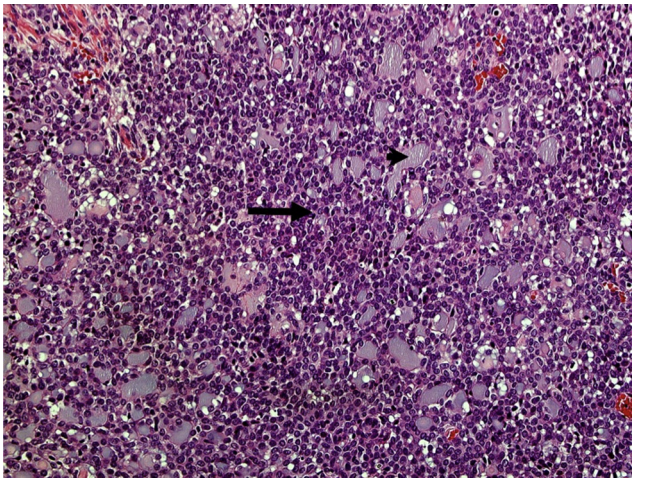

Figure 3. Malignant basaloid cells with hyperchromatic nuclei and sparse cytoplasm in nests (arrow) and multiple, adjacent pseudocystic structures (arrowhead) characteristic of the cribriform pattern of adenoid cystic carcinoma (hematoxylin-eosin, 200×).