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Advanced Ethmoid Sinus Adenocarcinoma Presenting as Temporary Exudative Retinal Detachment

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Key Words
Adenocarcinoma · Ethmoid sinus · Temporary exudative retinal detachment

Abstract
We describe the case of a 70-year-old male patient with ethmoid sinus adenocarcinoma who developed an exudative retinal detachment (ERD) in the right eye as the first manifestation. Two weeks after presentation, total regression of the ERD was noted. Extensive investigations for local causes of ERD were unrewarding. Finally, we performed a computed tomography scanning of the head that revealed an ethmoidal mass extending to the orbit. The diagnosis of adenocarcinoma was confirmed by biopsy. Neoplastic phenomena should be considered in patients presenting with temporary ERD.

Introduction

The annual European incidence of nasal cavity cancers is no more than 2 per 100.000 in men and less than 1 per 100.000 in women, while the 5-year relative survival in the period between 1990 and 1994 was 45% [1]. Ethmoid sinus cancers account for 10% of paranasal sinus cancers and 1% of all malignant head and neck tumours [1]. Adenocarcinoma constitutes the prevailing histological type among ethmoidal malignancies, with a reported frequency of 60–75% [2]. The most common presenting symptoms include proptosis, diplopia, visual loss, nasal obstruction and/or discharge, facial oedema and local pain [2, 3]. Exudative retinal detachment (ERD) is usually caused by intraocular inflammation but may

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be associated with malignancy. Ethmoidal carcinoma has not previously been reported to be associated with ERD.

Case Report

A 70-year-old Caucasian man was referred with a 2-week history of reduced vision in the right eye. He had no past ocular or medical history and reported no systemic or neurologic symptoms. On examination, corrected visual acuity was 1/10 in the right eye and 20/20 in the left eye. Ocular motility and the anterior segment examination were normal. There was no proptosis. Intraocular pressures were 14 mm Hg in the right eye and 12 mm Hg in the left eye. The mode B ocular ultrasound detected a localized ERD involving temporal superior (fig. 1a). Examination of the posterior segment of the left eye was unremarkable.

Two weeks after presentation, the patient reported mild pain, and no vision in the right eye. On examination, visual acuity was no light perception in the right eye and 20/20 in the left eye. A right relative afferent pupillary defect and an external deviation of the right globe were noted (fig. 1b). The ERD had completely resolved (fig. 1c), and mottling of the retinal pigment epithelium was evident. Computed tomographic scan of the brain and orbits demonstrated a space-occupying lesion in the right ethmoid (fig. 2). An excisional biopsy of the ethmoid lesion revealed an adenocarcinoma (fig. 3). After stereotactic radiotherapy and chemotherapy, tumour recurrence occurred during the 10 months of follow-up. The patient died 2 months later.

Discussion

Malignant ethmoid tumours are rare sinonasal tumours [4] that include squamous cell carcinoma, adenocarcinoma and aesthesioneuroblastoma. Although squamous carcinoma is the predominant histological type in other paranasal neoplasms, adenocarcinomas constitute 60–75% of ethmoidal cancers [2], and they are associated with a worse prognosis, with survival rates of only 40% at 3 years and 0% at 5 years for T4 tumours (in our case, the tumour extended from the right ethmoid sinus to the orbit) [5, 6]. Epidemiologically, adenocarcinomas have been associated with workers exposed to hardwood dust, chromium, nickel-refining processes, and leather tanning [7]. Direct spread of an ethmoid sinus carcinoma through the orbit is not unusual. In contrast, temporary ERD as the first manifestation of an advanced ethmoid adenocarcinoma has not been reported. The ocular presentation of ERD is usually associated with direct invasion. This is a rare case of ethmoid sinus carcinoma presenting initially as ERD. A search of the literature (MEDLINE) revealed no previously reported cases. In this patient, direct invasion was unlikely because no direct invasion was visible on imaging. ERD may develop as a result of choroidal involvement by malignant cells causing retinal pigment epithelial disturbances or due to incompetence of the outer blood retinal barrier inducing retinal pigment epithelial changes. Decreased visual acuity, a presenting symptom in 27% of cases with secondary orbital affectation [8], did not contribute to the early diagnosis of this cancer, especially in absence of other symptoms such as nasal congestion and epistaxis. Any tumour with an intranasal component can be approached endoscopically, thus allowing histologic confirmation [3]. If the diagnosis of a malignant sinus tumour is confirmed, palliative treatment is advocated for patients with advanced disease. In the world's largest published series, no patient survived longer than 10

months after surgery [9]. For patients with such advanced disease, radiotherapy and adjuvant chemotherapy are good palliative treatment methods [3, 10].

This case highlights the possibility that ethmoid adenocarcinoma may present as an ocular pathology in the absence of signs or nasal symptoms. Extensive investigations for local and systemic causes of ERD must be performed. Clinical ophthalmologists should be aware that in rare cases with orbital involvement, visual affectation might mask the diagnosis of a malignant sinus disease.

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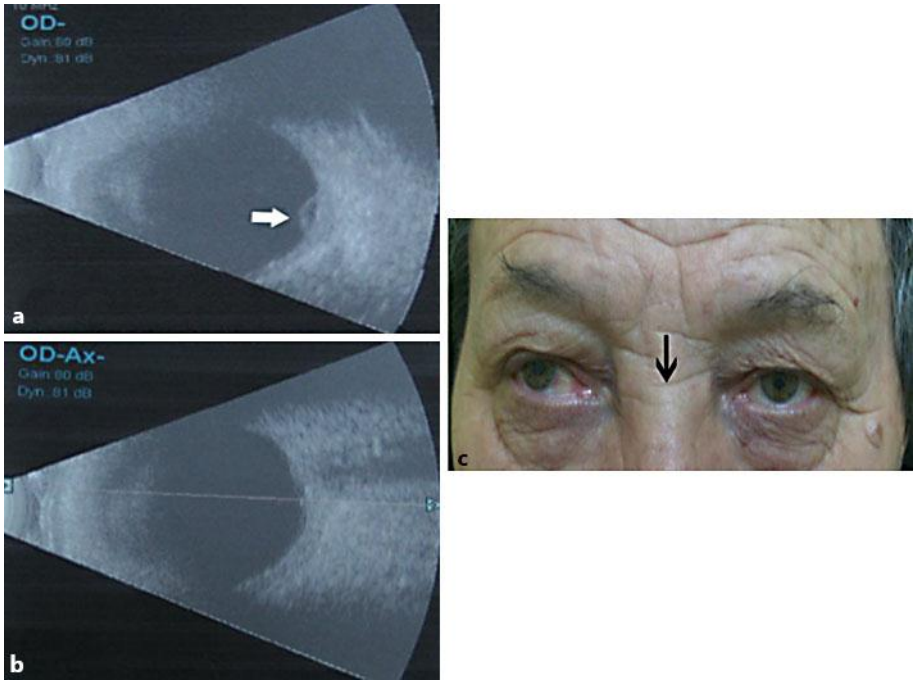

Fig. 1. **a** B scan image showing a smaller ERD (white arrow). **b** B scan image after 2 weeks of follow-up showing a resolved retinal detachment. **c** External appearance of the fronto-orbital region of the patient showing tumoral dilatation of the nasal bridge (black arrow).

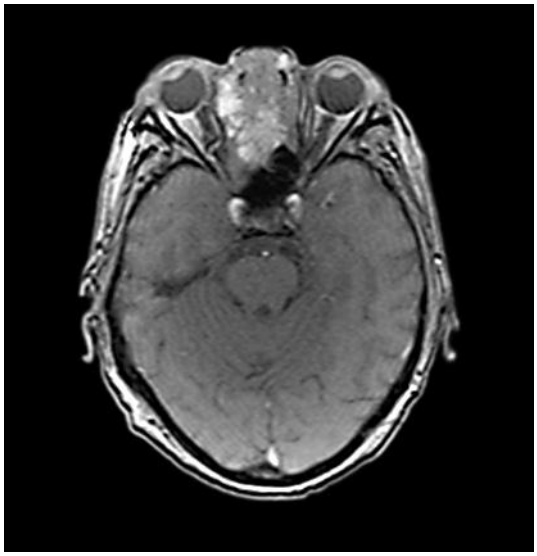

Fig. 2. Axial computed tomography showing a rounded heterogeneous mass in the right ethmoid sinuses extending through the lamina papyracea into the orbit.

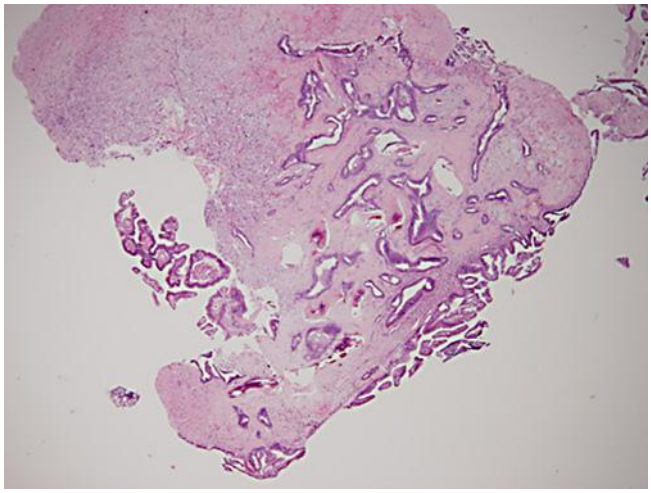

Fig. 3. Endoscopic biopsy of the right frontal ethmoid sinus revealed an adenocarcinoma resembling a colonic adenocarcinoma with back-to-back glands of pleomorphic columnar cells with intracellular mucin, but no prominent goblet cells (HE ×100).