

**Case report** 

Immunological Disorders and Immunotherapy

# Bilateral Choroidal Metastasis as Initial Presentation of Parotid Gland Adenoid Cystic Carcinoma

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#### Abstract

Choroidal metastasis rarely reveals a primary cancer. At this stage, they are associated with multiple metastases. Adenoid cystic carcinoma also known as cylindroma is very rare and constitutes 1% of major and minor salivary glands malignancies. In the parotid gland, the ACC is relatively rare, constituting only 2–3% of all tumors. The purposes of this study are to report the first case of parotid gland adenoid cystic carcinoma metastatic to the choroid and discuss treatment approaches. The systemic prognosis of salivary gland ACC is poor, with frequent local recurrences and late distance metastases. Even though uncommon, ocular metastases should be considered, and plaque radiotherapy may be useful to treat these patients.

Keywords: Choroid; Metastasis; Parotid gland; Adenoid cystic carcinoma

### Introduction

The choroid accounts for around 90% of metastasis to the uveal tract. In 1997, Shields et al. reported personal experience with a series of 520 eyes from 420 patients with uveal metastasis; the most common primary sites were breast (40%), lung (36%), and skin melanoma (8%). There were no cases of unveil metastasis from parotid gland carcinoma in the series of 420 patients [1].

Adenoid cystic carcinoma (ACC) is the second most common malignancy of the salivary glands after mucoepidemoid carcinoma [2].

Although rare, ACC has been previously documented to metastasize to the choroid [3-6] and to the iris [7]. We herein report an unusual case of parotid gland ACC revealed by bilateral choroidal metastasis.

#### **Case Presentation**

A 52-year-old man, with no prior medical history, complained of decreased vision in his left eye for 1 week. Entering corrected acuities were 10/10 in the right eye and hand motion in the left.

Anterior segment findings were unremarkable and intraocular pressures were 11 mmHg in the right and 12 mmHg in the left.

The right and left fundi are shown. The right fundus demonstrated serous retinal detachments. The left fundus showed an elevated serous retinal detachment of the posterior pole with macula off (Figures 1 and 2).

B-scan ultrasonography reveals an elevated choroidal tumor at temporal area more marked in the left eye (Figures 3 and 4).



Figure 1: Fundus photo of right eye showing serous retinal detachments.

Clinical examination of the patient revealed a right peripheral facial palsy with a right retro auricular swelling (Figure 5).

The ultrasonography findings indicated a mass suggestive of a tissue consistency located at the right parotid.

Magnetic resonance imaging showed a nodular mass in the right parotid gland locally invasive with cervical extension, and secondary location in the brain and the choroid (Figure 6).

Chest radiography showed canon-ball metastases. Abdominal and Pelvic CT and bone scan were normal.

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**Figure 2:** Fundus photo of left eye showing retinal detachment with macula off.



Figure 3: B-scan ultrasound of right eye.

Our patient underwent parotidectomy. The diagnosis was consistent with adenoid cystic carcinoma of the parotid gland with brain and choroidal.

The patient received palliative treatment combined chemotherapy with 5 fluorouracil and cisplatin and whole-brain irradiation therapy.

## Discussion

The parotid glands are the largest of the major salivary glands. ACC has the highest incidence of distant metastasis among carcinomas of the parotid gland, occurring in 30%–50% of patients by 20-year follow-up. The most common distant metastatic sites include lung, bone, and central nervous system [2].



Figure 4: B-scan ultrasound of left.



**Figure 5:** 52-year-old man with swelling right peripheral facial palsy with a right retro auricular swelling of the parotid.

According to our literature search, there have been only four single case reports of choroidal metastasis from ACC of the salivary glands in the ophthalmic literature. One case reported of iris metastasis from parotid gland ACC.

Jenrette and Fitzgerald reported a 57-year-old woman with ACC of the submandibular salivary gland who developed choroidal metastasis 11 months after diagnosis [3]. Shields et al. described a 51-year-old woman with choroidal metastasis from an ACC of the submandibular salivary gland 14 months after diagnosis [4]. Demirci et al. described a 50-year-old woman with ACC of the submandibular salivary gland who developed choroidal metastasis 5 years after diagnosis [5]. Gutmann et al. reported an 88-year-old man with choroidal metastasis from ACC of the submandibular salivary gland, who was treated with enucleation for suspected choroidal melanoma [6]. Montero J et al. described a 64-year-old man with iris metastasis from an ACC of the parotid gland 5 years after diagnosis [7].

All the choroidal metastasis reported the primary site in the submandibular salivary gland, and our case is the first with primary site in the parotid gland.

Treatment alternatives for uveal metastasis include observation if end stage, excisional biopsy for isolated metastasis, and radiotherapy or chemotherapy for patients with multiple metastases.



**Figure 6:** Magnetic resonance imaging showed a tumor in the right parotid gland with secondary location.

Radiotherapy is the most commonly used modality as it will provide local control and help to avoid visual loss, pain and morbid enucleation especially in tumors with large size and subretinal fluid. Plaque radiotherapy can be used if tumor is small and located away from macula and optic nerve and in patients refractory to radiotherapy as high dose can be provided. Response can be evaluated 6-8 weeks post-treatment [8].

To date, radiotherapy remains the treatment of choice of choroidal metastases: long experience and good results are reported in the literature [9].

From the above-cited 5 cases, 3 were treated with external beam radiotherapy, 1 case was treated with plaque radiotherapy and 1 case

was treated with enucleation. Our patient was treated with combined chemotherapy and radiotherapy.

The systemic prognosis of salivary gland ACC is poor, with frequent local recurrences and late distant metastases. The 5-year survival rate after effective treatment is 75%, but long-term survival rates are low (10 years – 20% and 15 years – 10%) [10].

## Conclusion

ACC is a rare malignant tumor of the parotid gland. Metastasis can manifest very late, and hence a long-term follow-up and a high index of suspicion are necessary to diagnose it early in order to enable a more favorable prognosis and better quality of life. Even though uncommon, ocular metastases should be considered in these patients, and periodic eye examinations are advised.

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