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Primary Adenoid Cystic Carcinoma Presenting as an Orbital Apex Tumor

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Abstract

Primary adenoid cystic carcinoma occurring in the orbital apex is rare. We present the clinical features of a patient who initially presented with the clinical and radiologic features of an orbital pseudotumor. He developed features of orbital apex syndrome and repeat imaging showed a tumor of the orbital apex with intracranial invasion. He underwent radical skull base surgery and pathologic examination revealed adenoid cystic carcinoma in the orbital apex with a normal lacrimal apparatus. He received post operative radiation and the outcome in the light of a review of available literature is being discussed.

Keywords

Adenoid cystic carcinoma; orbital apex; extra lacrimal; intracranial

INTRODUCTION

Adenoid cystic carcinoma is a slow growing, locally invasive tumor of epithelial origin.[1,2] In the head and neck, the common sites of origin are the minor and major salivary glands and also the lacrimal glands. Adenoid cystic carcinoma accounts for 4.8% of primary orbital neoplasms and commonly arises superolaterally from the main lacrimal gland, and rarely may arise in the medial aspect of the orbit from the lacrimal apparatus.[3–6] Primary adenoid cystic carcinoma of the orbit from an extra lacrimal region is quite rare.[6] We report a unique presentation of an adenoid cystic carcinoma, presenting as an orbital apex mass, mimicking a benign lesion and subsequently showing features of intracranial infiltration. The radiologic features, management and the outcome in the light of a review of available literature is being discussed

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CASE REPORT

A 51 year old gentleman of Indian ancestry presented to the regional cancer center in October 1998, with a history of proptosis, progressive deterioration of visual acuity of the left eye, diplopia and left hemicranial headache of two years duration. There was no history of previous trauma or any other intercurrent illness. He had been seen at different hospitals earlier, investigated and managed with analgesics and steroids.

His initial complaints were progressive painless proptosis and worsening of visual acuity of the left eye. CT scan of the orbit done a year previously, in august 1997, had shown a well defined irregular hyperdense mass lesion in the apical and medial aspect of the left orbit which showed irregular contrast enhancement. The lacrimal gland and apparatus were normal. There was no bony involvement. Moderate proptosis was noted. T1 weighted MRI scan of the orbit revealed a well defined hypointense retrobulbar mass lesion which was hypointensive on T2 weighted also. There was no significant increase in signal intensity in the T1 weighted images after gadolinium contrast. The mass was in the medial extraconal location involving the orbital apex. The optic nerve appeared free (Figure 1).

The clinical and radiologic appearance were similar to a benign lesion, probably an orbital pseudotumor or a granulomatous lesion and no attempt at a histologic diagnosis was made at that time. He received steroids with partial improvement of symptoms which worsened after a year when he developed progressive loss of visual acuity and diplopia. CT scan was repeated in October 1998, which showed that the hyperdense extraaxial mass lesion had extended through the superior orbital apex to the parasellar region *and* was involving the left cavernous sinus wall. MR image showed a mass lesion of the orbital apex and retrobulbar region appearing hypointense on T1 WI and T2 WI with moderate increase in signal intensity on gadolinium contrast images. It was found to involve the posterior part of the extraocular muscles and extending posteriorly into middle cranial fossa up to cavernous sinus and the posterior part of the Merckel's cave.

Infiltration of the parapharyngeal muscles was present inferiorly. Cavernous segment of ipsilateral internal carotid showed normal flow void. The patient had complete loss of vision of the left eye within two months, when he was seen by us. Clinical examination including complete neuro-ophthalmologic examination showed ptosis of the left eye and proptosis of 22 mm and a dilated pupil with no reaction to light. Corneal reflex was sluggish. He also had complete loss of left ocular movements and sensory deficit over the maxilla. The III, IV, and VI cranial nerves were affected on the left. Papilloedema was present. Thorax and abdomen were normal on imaging.

A left medial orbitotomy and biopsy was carried out. Histopathology of the tumor showed a malignant neoplasm infiltrating muscle tissue having a cribriform appearance. Neoplastic cells had a round vesicular nucleus and scanty cytoplasm (Figure 2). The picture was that of an adenoid cystic carcinoma showing characteristic perineural infiltration in certain areas. The patient underwent an orbital exenteration. Peroperatively the retroorbital tumor was found to involve the ocular muscles and optic nerve, with parasellar and cavernous sinus extension which encased the internal carotid artery. The lacrimal gland and sac were free of tumor. Skull base surgery with near total resection of the intracranial extension was carried out. There was no detectable primary malignancy elsewhere. He received postoperative external radiation to the primary site to dose of 50 Gy in 25 Fractions. Patient is still alive with no evidence of disease.

DISCUSSION

Adenoid cystic carcinoma, first described by Billroth as cylindroma, is a tumor of epithelial origin. In the head and neck it usually arises from the salivary glands, in the orbit from the lacrimal glands and rarely from mucous glands of other sites.[1,2] The common orbital presentation is that of a palpable mass in the superior temporal quadrant. Associated painful proptosis and inferomedial displacement of the eyeball may be seen. Clinical course of the tumor is of progressive bony erosion of the orbital wall, contiguous growth along the roof to the orbital apex and perineural infiltration with intracranial spread.[1,2]

Adenoid cystic carcinoma is known for its subtle presentations and rarely adenoid cystic carcinoma of the orbit may arise from sites other than the lacrimal gland. Three cases of adenoid cystic carcinoma in the medial aspect of the orbit, arising from the lacrimal apparatus have been reported.[3–5] Shields et al. have reported a case of adenoid cystic carcinoma, of possible ectopic lacrimal gland origin, arising in the anteronasal region of the orbit.[6]

The orbital apex syndrome includes the third , fourth and sixth cranial nerve deficits. Sometimes involvement of the first and second divisions of the fifth nerve, ocular sympathetic paralysis, proptosis, loss of vision and conjunctival chemosis may also be associated findings. Differential diagnosis of lesions causing orbital apex syndrome include inflammatory pseudotumors, trauma, neural tumors, meningiomas, lymphoid tumors, vascular tumors, carcinomas, metastatic lesions, thyroid orbitopathy, mycosis, sarcoidosis, and other granulomatous disorders.

A case of adenoid cystic carcinoma arising from the paranasal sinus with classical orbital apex syndrome as the primary presentation has been reported by McDonald.[7] Lee et al. have reported a patient in whom the tumor mimicked an orbital pseudotumor with subsequent development of intracranial involvement possibly by contiguous perineural spread.[8] Tse et al. have reported metastatic adenoid cystic carcinoma from a lacrimal gland primary presenting with orbital apex syndrome.[9] Though clinically similar to this presentation, Tse et al. could not process the complete lacrimal gland in their study and did in fact detect a possible occult primary adenoid cystic carcinoma originating in the lacrimal gland, and hypothesize that carcinoma cells may have escaped from the lacrimal gland in the early phase of carcinogenesis.

We could not find any abnormality in the lacrimal gland even after serial sectioning, suggesting a possible occult extralacrimal origin of the adenoid cystic carcinoma. Similar to our experience, the case report by Tse et al. also underlines the importance of multi-modal treatment in orbital adenoid cystic carcinoma. Primary presentation of intracranial adenoid cystic carcinoma mimicking meningioma has also been reported previously.[10,11]

The possibility of origin of adenoid cystic carcinoma from ectopic lacrimal gland tissue has been proposed by Shields.[6] There are also reports of benign mixed tumors and of microscopic focus of adenocarcinoma in ectopic lacrimal gland tissue.[12,13] There have been reports of inflammation of retroorbital ectopic lacrimal gland tissue presenting with axial proptosis.[12]

In this report, the presenting complaint of the patient was axial painless proptosis without any displacement of the globe. The mass lesion at presentation had no signs of bony or neural infiltration and the diagnostic possibilities thought of were benign lesions like orbital pseudotumor or a granulomatous disease like sarcoidosis or infectious myosistis. CT scan in a case of adenoid cystic carcinoma may show the characteristic hyperdense mass with irregular serrated borders which enhances homogenously with contrast, with perineural and extraocular muscle invasion, extension medially and posteriorly with bone erosion and destruction.[14]

MRI appearance of an adenoid cystic carcinoma is that of an irregular well defined infiltrating mass with heterogenous hyperintense signal compared to extraocular muscles with hypointense signal compared to orbital fat on T1 weighted images. On T2 weighted images it may be heterogeneously hyperintense compared to extraocular muscles and orbital fat. Calcifications may be present which appear hypointense on both T1 and T2 weighted images. Marked focal enhancement on Gadolinium contrast is commonly seen.[14,16] A benign orbital inflammatory lesion may be isointense to hyperintense to extraocular muscles and hypointense to fat on TI weighted images and hypo, isointense or hyper intense on T2 weighted images.[14–16]

In our patients the mass lesion presented in an unusual site , with no signs of bony or neural infiltration. The mass on MRI scan was hypointense with no significant increase in signal intensity on contrast and was hypointense on T2WI. The pitfall of making a benign diagnosis on imaging alone and managing conservatively is amply demonstrated by this report. The patient showed improvement with steroids which delayed a pathologic diagnosis. The ideal option would have been definite pathologic diagnosis initially or if alternatively a non granulomatous inflammatory lesion is suspected, periodic ultrasound orbit, CT or MR scans and immediate surgical intervention when the tumor did not respond to medical management and before it showed the slightest signs of invasion and intracranial extension.[17]

Adenoid cystic carcinoma has high rates of local recurrence and distant metastasis with three fourth of the patients relapsing on long term follow up.[2,7] Orbital exenteration has been recommended for adenoid cystic carcinoma, as perineural and perivascular spread within skull base foramina and intracranial extension is common.[2,7,18] Preservation of the carotid artery and a complete resection of the cavernous sinus preserving the third and fourth cranial nerves along with an orbital exenteration may be attempted to allow the patients to have long term survival with optimal functional and acceptable cosmetic results.[18]

Tse et al. have suggested the utility of pre-operative intraarterial chemotherapy if intracranial extension is present, to facilitate exenteration of adenoid cystic carcinoma.[9] Even after en bloc exenteration or cranio-orbital resection, the chances of local recurrence are quite high and post operative radiation has been shown to decrease the local recurrence rates.[19] The long term prognosis for adenoid cystic carcinoma, even after multi-modal treatment with radical surgery and local radiotherapy or chemotherapy, has been poor in most reported studies.[2,7,18,19]

The patient in this report did not have any detectable primary tumor elsewhere and the lacrimal gland and sac were normal. Further the tumor had a subtle presentation resembling the clinical and radiologic features of a nonmalignant lesion and then showed the classical features of slow relentless contiguous progression with intracranial infiltration associated with a malignant neoplasm. In such a situation, though rare, one could consider the possibility of adenoid cystic carcinoma arising from ectopic lacrimal gland tissue. This case illustrates the importance of a pathologic diagnosis and early intervention in similar presentations.

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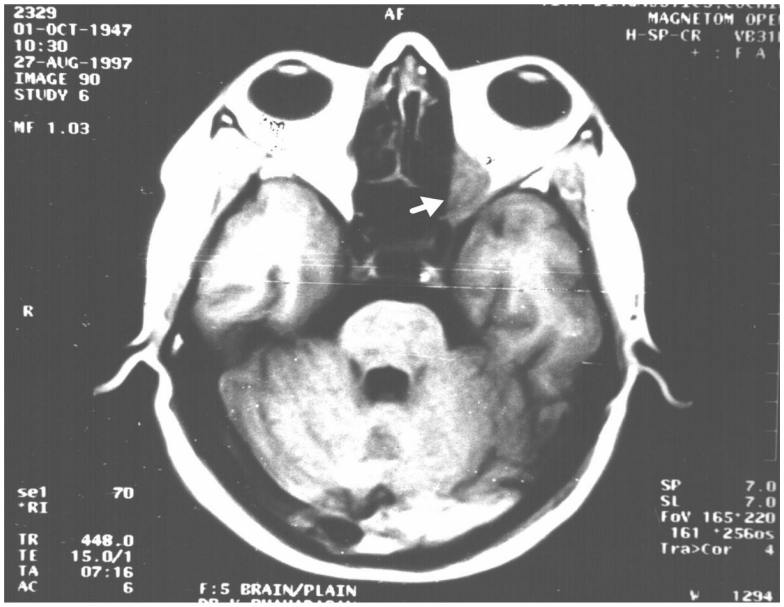

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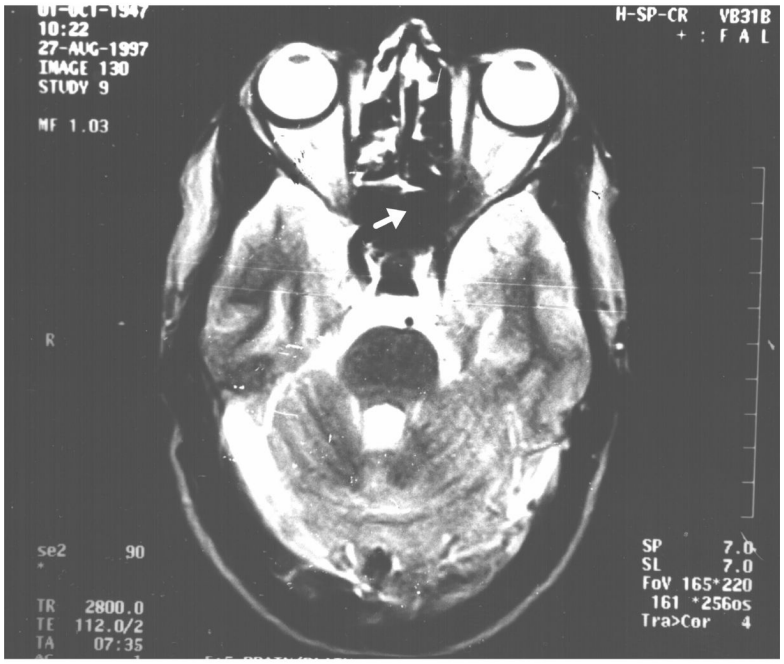

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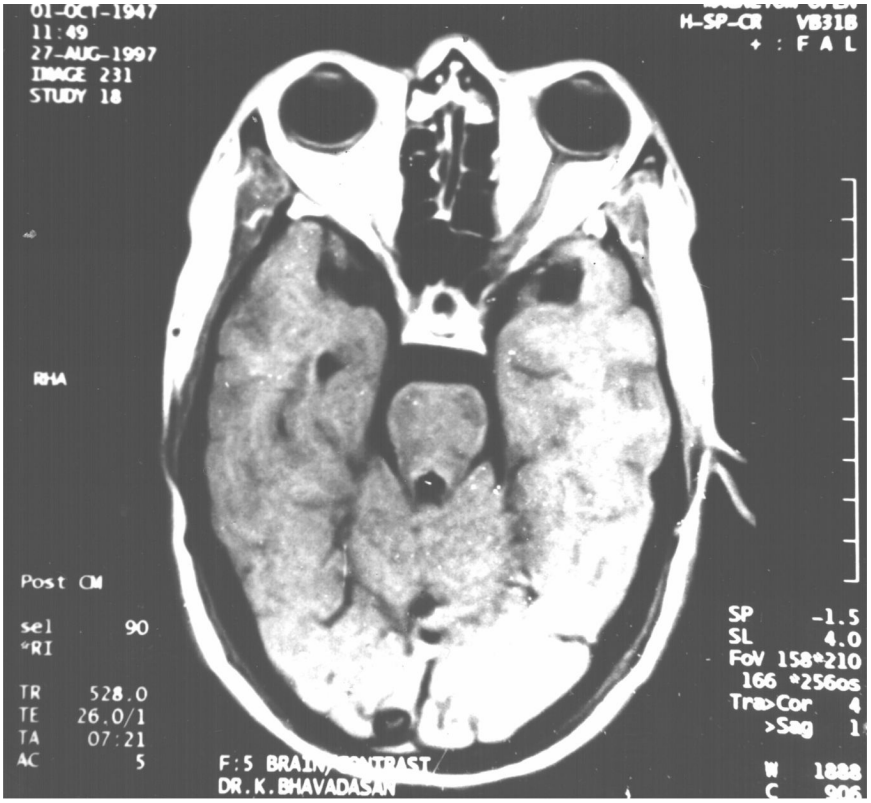
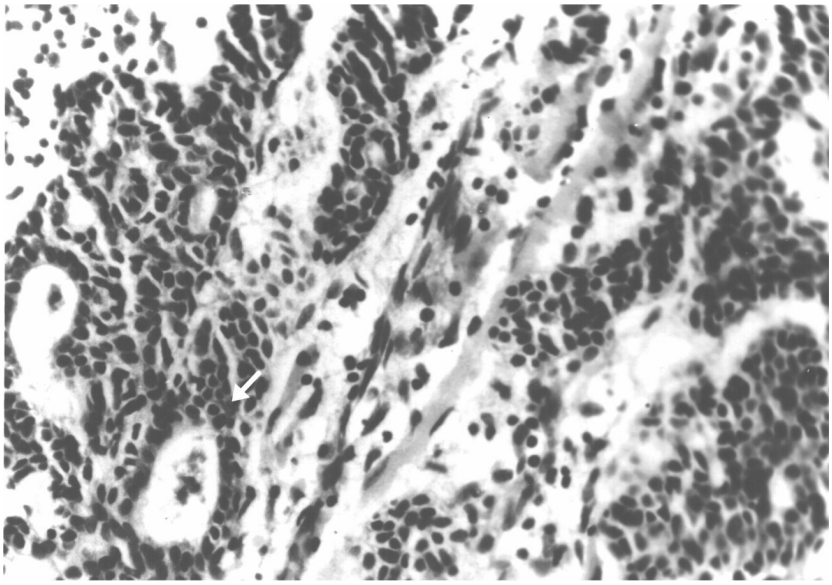


(C)

FIGURE 1.

A. T1 weighted MR image showing the irregular lesion with low signal intensity in the apex of the left orbit (dated 27.08.1997); B. T2 weighted MR image showing the hypointense mass lesion; C. T1 weighted MR image with Gadolinium contrast shows no significant increase in signal intensity. Optic nerve is intact. *(Continued)*



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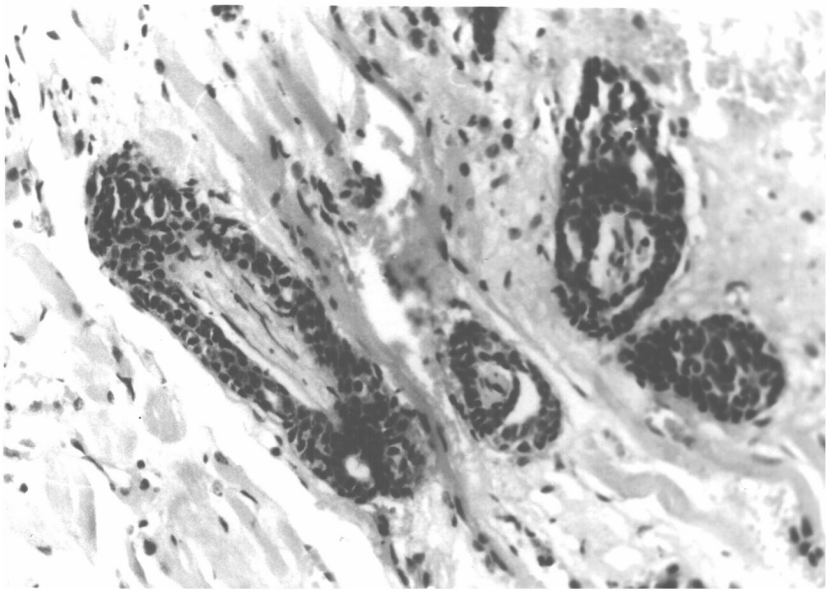

(B)

FIGURE 2.

A. Histopathologic examination of the resected orbital apex tumour showing adenoid cystic carcinoma infiltrating muscle—characteristic cribriform pattern of arrangement of myoepithelail cells around cystic spaces seen on the left (H & E section X 450); B. The resected specimen showing characteristic perineural infiltration of adenoid cystic carcinoma.