A 32 year-old Hispanic man was referred with two months of progressive left upper lid "fullness" and ptosis OS. At the onset of his symptoms, he consulted a physician who treated him with antibiotic eye drops without relief. One month prior to referral, he noted "blurry vision" OS as well as binocular vertical diplopia in upgaze. He denied ocular pain.

Review of systems was remarkable for mild right facial weakness and numbness as well as a "funny feeling" in his left hand during the previous four months. He complained of decreased memory and difficulty with mathematical calculations throughout the previous year. He had no systemic symptoms. Past medical history was unremarkable.

On examination, visual acuity was 20/20 OD and 20/30 OS. External exam was remarkable for left upper lid fullness and hypoglobus OS. There was 3mm of proptosis and resistance to repulsion OS. Pupils were equal and reactive OU with no RAPD. Ocular motility revealed full ductions OD and 60% of normal elevation OS. Goldmann visual fields were full OU. Fundus exam revealed pink/flat optic nerves OU and choroidal folds in the superior fundus OS. Neurologic exam was remarkable for decreased sensation in a right CN V1/V2 distribution; a right peripheral CN VII palsy; mild weakness and decreased sensation to light touch of the left upper and lower extremities and agraphesthesias of the left hand. He had mild tenderness over the right mandibular angle and an enlarged, non-tender right cervical lymph node.
Title of Presentation: Here, there and everywhere

Authors: Raul A. Guevara, MD, Paul H. Phillips, MD, Nancy J. Newman, MD

Final Diagnosis: Adenoid cystic carcinoma

Summary of case including pathology and references (200-300 words):

MRI (brain): 4 cm extraaxial mass in the right parietal region with meningeal enhancement, minimal surrounding edema, significant mass effect and midline shift. There was a second mass in the left superior orbital ridge encroaching on the left globe with inferior displacement.

CT (brain and orbits): The above lesions were associated with lytic bone lesions. The lytic lesion in the right calvarium appeared to be the origin of the intracranial mass. There was a lytic lesion in the right mandibular angle. The orbital mass was associated with contiguous bone destruction and extended into the the cranium.

CT (thorax, abdomen and pelvis): No evidence of metastases.

Bone Survey: Lytic lesions of the right skull and right mandible.

Bone scan: Increased uptake in the left orbit, right mandible and midline skull.

Labs: CBC, Chem-18, SPEP, UPEP - normal

Biopsy of the right parietal mass: Adenoid Cystic Carcinoma

Course: The patient was discharged with the plan to pursue palliative radiation treatment and chemotherapy.

Adenoid cystic carcinoma may originate in a variety of anatomic sites including the lacrimal glands, the major and minor salivary glands, the parotid gland, the mucous glands of the upper respiratory and digestive tracts, as well as the breast, cervix and skin. In our patient, the left lacrimal gland is the most likely primary site. However, the right salivary gland may be the primary site as our patient had a right CN VII palsy and a lytic lesion in the right mandibular angle. Although no discrete mass was noted on neuroimaging of this region in our patient, Cummings described two cases of adenoid cystic carcinoma in which local signs occurred 4-6 years prior to the development of a clinically detectable parotid mass.¹

Adenoid cystic carcinoma is the most common malignant epithelial tumor of the lacrimal gland comprising 29% of all lacrimal epithelial neoplasms.² It commonly presents with orbital pain, mild proptosis and diplopia associated with enlargement of the gland. The pain occurs secondary to perineural invasion and bone destruction. Patients typically present within 10 months of onset of symptoms.² The tumor is locally invasive and infiltrates nerves and blood vessels. The clinical course is characterized by multiple local recurrences despite attempts at surgical resection. Although treatment frequently involves exenteration of
PROGRAM FORM (ANSWER)

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the orbit with removal of affected bone and post-operative radiation, this has not been demonstrated to increase survival. The prognosis is poor with approximately 50% survival 21/2 years after diagnosis.

The frequency of intracranial invasion of adenoid cystic carcinoma is between 4% and 22%. This usually occurs by direct tumor invasion or via perineural spread along cranial nerves with involvement of the skull base. Hematogenous metastases, when they occur, are usually to the lung, and CNS metastasis is rare. Gelber et. al. described a patient with adenoid cystic carcinoma of the parotid gland with a histologically confirmed metastatic lesion to the inner table of the left frontal calvaria, epidural compartment, and brain parenchyma. Variakojis et. al. described a patient with adenoid cystic carcinoma arising from the right submaxillary salivary gland with a metastatic lesion to the left frontoparietal region of the skull which invaded the underlying dura and brain. Hara et. al. reported a patient with an isolated adenoid cystic carcinoma of the frontal lobe from an unknown primary site. These metastatic CNS lesions occurred in anatomic positions inconsistent with direct or retrograde perineural spread from the primary site. Our case likely demonstrates hematogenous CNS metastatic disease from a lacrimal gland adenoid cystic carcinoma.

REFERENCES: