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Adenosquamous Carcinoma of the Facial Bones, Skull Base, and Calvaria: CT and MR Manifestations

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Summary: A 62-year-old woman had proptosis of the right eye, decreased visual acuity of the left eye, and no other focal neurologic deficits. She had had a grand mal seizure 1 month before admission. The CT and MR studies showed extensive bone destruction of the margins of the right orbit, the floor of the middle cranial fossa, the right cavernous sinus, and much of the calvaria. There was considerable dural disease and tumor in the right orbit, paranasal sinuses, and scalp, as well as mucocoeles of the left ethmoidal sinus with desiccated secretions. The diagnosis was adenosquamous carcinoma, an aggressive tumor related to both squamous cell carcinoma and adenocarcinoma.

Index terms: Carcinoma; Skull, neoplasms

There is an old adage that uncommon presentations of common things occur more commonly than common presentations of rare things. Although it is probably useful to be aware of such adages, occasionally one does encounter the rare case and it is usually the pathologist who establishes the diagnosis. We report here our findings in a 62-year-old woman who presented with unilateral proptosis and a paucity of other disorders, yet who had extensive adenosquamous carcinoma that on computed tomographic (CT) scans and magnetic resonance (MR) images was seen to have caused massive destruction of the facial bones, calvaria, and skull base, with tumor in the orbit, paranasal sinuses, cavernous sinus, scalp, and dura.

Case Report

A 62-year-old woman had slowly progressive proptosis of the right eye, which first became evident 4 months before admission. In other respects, the patient remained healthy. One month before admission she had a grand mal seizure, after which she sought medical attention. She had no other significant medical history. On physical examination, there was marked proptosis of the right eye, slight

fullness of the right side of the face, and a right-sided nasal mass. There was a full range of extraocular muscle movement; however, her visual acuity in the left eye was to light perception only. With this exception, there were no neurologic deficits and she had neither headache nor scalp or facial pain. A contrast-enhanced CT scan showed extensive bone destruction of the right facial bones, the floor of the right anterior and middle cranial fossae, the right cavernous sinus, and the calvaria, involving most of the right side as well as the upper portion of the left side (Fig 1A–E). Tumor was present both superficial and deep to the destroyed calvaria and there was dural thickening in these areas. Much of the involved bone was thicker than normal, suggesting a blastic change. Multiple expansile masses were present in the left ethmoidal complex, possibly representing mucocoeles. Tumor extended extraconally into the right orbit, causing proptosis that stretched the optic nerve to its extreme. The right-sided nasal mass had bone overlying its anteroinferior margin, possibly representing residual turbinate involved with tumor. A 2-cm partially necrotic mass was present in the right parotid gland, presumably representing a metastatic node. Contrast-enhanced MR imaging of the head and facial region confirmed the distribution of the disease as seen on CT scans (Fig 1F and G). The invasion of the right cavernous sinus and the more extensive degree of dural disease was more evident on the MR images. Also better seen was the amount of tumor at the orbital apex spreading extraconally into the orbit. The masses in the left ethmoidal complex and in the right nasal cavity had varying signal intensities on T1-weighted and T2-weighted MR images that were consistent with mucocoeles containing partially desiccated secretions. The signal intensities of these secretions differed from those of the tumor, which had intermediate T1-weighted and T2-weighted signal intensities. The tumor also had fairly intense enhancement on contrast-enhanced MR images.

Two biopsies of the nasal mass were performed; however, they revealed only bone and inflammatory tissue. Finally, a biopsy specimen of the left side of the scalp showed adenosquamous carcinoma (Fig 1H). Because of

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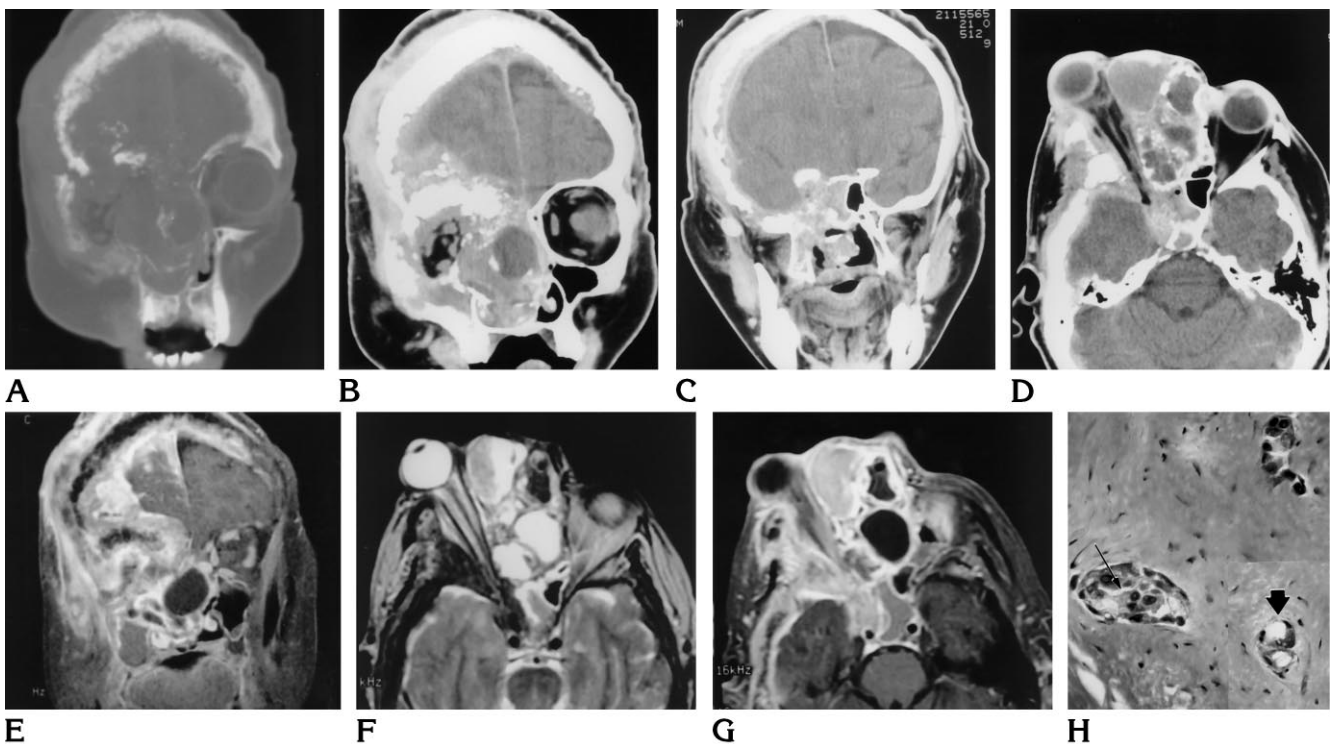


Fig 1. Sixty-two-year-old woman with proptosis of the right eye and decreased visual acuity of the left eye.

A and B, Coronal wide window (A) and narrow window (B) settings of a CT scan of the facial region show massive destruction of the nasal structures, the bony margins of the right orbit, the right maxilla, and much of the calvaria. In B, tumor can be seen surrounding the orbit and lying both superficial and deep to the calvaria.

C, Coronal CT scan through the sphenoidal and cavernous sinus region shows tumor destroying the right sphenoidal sinus, cavernous sinus, upper pterygoid process, and calvaria. The right dural thickening can also be seen.

D, Axial CT scan shows multiple masses in the ethmoidal complex, suggestive of mucocoeles. Tumor is seen in the right sphenoidal sinus, cavernous sinus, the dura about the right middle cranial fossa and orbital apex, and along the lateral margin of the right orbit. There is extreme proptosis of the right eye.

E, Coronal T1-weighted contrast-enhanced fat-suppressed MR image shows extensive tumor involving the right orbit and calvaria, with dural thickening. Tumor is both superficial and deep to the destroyed calvaria.

F, Axial T2-weighted MR image shows multiple fluid-containing masses in the ethmoidal complex, most of which had intermediate T1-weighted signal intensity. There is extreme proptosis, with stretching and thinning of the right optic nerve.

G, Axial T1-weighted contrast-enhanced, fat-suppressed MR image shows enhancing tumor in the right cavernous sinus, orbital apex, lateral orbital wall, and calvaria.

H, Photomicrograph shows islands of squamous cell carcinoma infiltrating dense fibrous tissue. Although keratinization is not seen here, fine intercellular bridges that are characteristic of squamous cell carcinoma can be seen (*thin arrow*). Inset shows glandular formation within tumor islands, probably as a result of discohesion between cells (*wide arrow*) (hematoxylin-eosin, original magnification $\times 400$).

the extensive nature of the tumor, the patient elected to leave the hospital without further treatment.

Discussion

Adenosquamous carcinoma and basaloid squamous carcinoma are related to both adenocarcinoma and squamous cell carcinoma. Because of their close relationship, they are discussed together. Adenosquamous carcinomas usually arise from minor salivary glands. The clinical presentation is usually different from that of squamous cell carcinoma in that the firm white to reddish exophytic mucosal tumor com-

ponent usually seen in squamous cell carcinoma is not present with either adenosquamous or basaloid squamous carcinomas. Instead, they are entirely submucosal, as are most salivary gland tumors. Occasionally, they may have an associated ulceration. Histologically, the adenosquamous carcinoma has the typical glands of adenocarcinoma, without the features of mucoepidermoid carcinoma, adenoidcystic carcinoma, or other malignant lesions of the salivary glands. The basaloid carcinoma has small cells, with a high nuclear-to-cytoplasmic ratio, that form glands with either a cribriform or

palisading appearance reminiscent of an adenoid cystic carcinoma or small cell-type carcinoma. However, the strong tendency of adenoid cystic carcinoma toward perineural spread is not present with these tumors. The squamous nature of these carcinomas may be indicated by a focal keratinization and/or pearl formation and/or a focal carcinoma in situ overlying the tumor. In this case, keratinization could be seen in addition to gland formation. The presence of focal overlying carcinoma in situ is suggestive of a diagnosis of squamous cell carcinoma, and such carcinoma in situ is not seen in the mucosa overlying other malignant lesions of the salivary glands. Often, examination of many histologic sections is necessary to identify carcinoma in situ, while at other times, one assumes that the in situ changes were abolished by the mucosal ulceration. The tumors so far reported in the literature have all been highly aggressive with dismal prognoses. It does appear that adenosquamous carcinoma and basaloid squamous carcinoma rarely present as either T1 or T2 tumors (1, 2).

The process of arriving at an imaging diagnosis is one of analyzing the findings and determining how, by their frequency of occurrence, they can best be assigned to a particular lesion. In the head and neck, between 70% and 90% of all the malignant lesions of the upper aerodigestive tract are squamous cell carcinomas. The variation in frequency is site-dependent, with nearly 90% of oral malignant tumors and about 80% of nasopharyngeal and sinonasal cavity tumors being squamous cell carcinomas (3–7). On CT scans, squamous cell carcinomas characteristically infiltrate and destroy adjacent bone (8). However, despite this aggressive imaging appearance, sinonasal squamous cell carcinomas uncommonly produce pain unless there is perineural invasion or an associated infection. The usual presenting disorder is nasal obstruction, and the average delay between onset of symptoms and final diagnosis is 6 months. As a result, these tumors can grow to considerable size by the time of diagnosis (7). Uncommonly, the tumor may chronically obstruct one or more paranasal sinuses, resulting in mucocele formation (9, 10). On MR images, squamous cell carcinomas usually have intermediate T1-weighted and T2-weighted signal intensity. These tumors have from slight to fairly intense enhancement on postcontrast MR im-

ages and, when large, focal regions of necrosis and hemorrhage may also occur (11).

Thus, in the present case, a diagnosis of squamous cell carcinoma at first seems appropriate on the basis of the frequency of the tumor and the presence of aggressive bone destruction. However, the unusually extensive degree of bone involvement in this case is not typical of squamous cell carcinoma. This is especially so when a presumed sinonasal tumor presents with such extensive calvarial and skull base bone destruction. The imaging appearance suggests that the tumor rapidly followed and destroyed the bone, extending well beyond the anatomic confines of the sinonasal cavity. In fact, the imaging appearance suggests that this tumor was behaving as an aggressive form of squamous cell carcinoma, and it seems appropriate that the diagnosis of rare adenosquamous carcinoma ought to be added to the imaging differential diagnosis in such aggressive-appearing cases.

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