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 mucoceles 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 mucoceles. 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 mucoceles 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
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 component 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

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 component 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 adeno-
id cystic carcinoma or small cell–type carci-
noma. 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 mu-
cosa overlying other malignant lesions of the
salivary glands. Often, examination of many
histologic sections is necessary to identify car-
cinoma in situ, while at other times, one as-
sumes 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 ap-
pear that adenosquamous carcinoma and basa-
loid squamous carcinoma rarely present as
either T1 or T2 tumors (1, 2).

The process of arriving at an imaging diag-
nosis is one of analyzing the findings and deter-
mining 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 aerodiges-
tive 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 char-
acteristically infiltrate and destroy adjacent
bone (8). However, despite this aggressive im-
ing 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 on-
set 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 ob-
struct one or more paranasal sinuses, resulting
in mucocele formation (9, 10). On MR images,
squamous cell carcinomas usually have inter-
mediate 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 ap-
propriate on the basis of the frequency of the tumor
and the presence of aggressive bone destruc-
tion. 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 squa-
mous cell carcinoma, and it seems appropriate
that the diagnosis of rare adenosquamous car-
cinoma ought to be added to the imaging dif-
fferential diagnosis in such aggressive-appear-
ing cases.

References

1. Gerughty RM, Henniger GR, Brown FM Adenosquamous carci-
noma of the nasal oral and laryngeal cavities: a clinicopathologic
2. Som PM, Brandwein M Salivary glands. In: Som PM, Curtin HD,
1996;2:894–895
3. Batsakis, JG. Squamous cell carcinomas of the oral cavity and
oropharynx. In: Tumors of the Head and Neck: Clinical and Patho-
logical Considerations. 2nd ed. Baltimore, Md: Williams & Wilkins;
1979:144–176
4. Batsakis JG. Carcinomas of the nasopharynx. In: Tumors of the
Head and Neck: Clinical and Pathological Considerations. 2nd ed.
Baltimore, Md: Williams & Wilkins; 1979:188–199
5. Sisson GA, Becker SP. Cancer of the nasal cavity and paranasal
Lawson W, Friedman WH, eds. Surgery of the Paranasal Sinus.
In: Tumors of the Head and Neck: Clinical and Pathological Con-
siderations. 2nd ed. Baltimore, Md: Williams & Wilkins; 1979:177–
187
8. Som PM, Shugar JMA The significance of bone expansion asso-
ciated with the diagnosis of malignant tumors of the paranasal
9. Som PM. Sinonasal cavity: Section Four, Tumors and Tumor like
Conditions. In: Som PM, Bergeron RT, eds. Head and Neck Imag-
10. Guerry RK, Smith JL. Paranasal sinus carcinoma causing orbital
and malignant sinonasal lesions with intracranial extension: their