Metastasis of soft-tissue myoepithelial carcinoma to clivus

Case report

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The clivus is involved in approximately 1% of intracranial tumors. The differential diagnosis for lesions involving the clivus includes tumor, infection, degeneration, and erosion. When considering expansile masses of the clivus, one should consider primary and metastatic tumors. Here we present the case of a myoepithelial carcinoma metastatic to the clivus as an aggressive tumor to consider in the differential diagnosis for clival masses when a history of myoepithelial carcinoma is present.

Case Report

History and Examination. This 14-year-old girl presented with left lateral gaze palsy with some left eye pain. She was otherwise neurologically intact. Her medical history was significant for a high-grade myoepithelial carcinoma of the thigh, first diagnosed 1 year previously. Following resection of the primary tumor, the patient was treated with proton beam therapy and chemotherapy (doxorubicin); her chemotherapy treatments were finished 7 months prior to presentation. In addition, 1 month prior to presentation, she was noted to have recurrent lung infections, with the workup revealing a metastasis to the left lung requiring resection. Ten days after surgery, the patient noted the above-mentioned sixth cranial nerve palsy. Neuroimaging demonstrated a 3 × 3–cm, osteolytic clival mass (Figs. 1 and 2).

Operation and Postoperative Course. A microscopic transsphenoidal subtotal resection with endoscopic assistance was performed. Pathological findings were consistent with the previously diagnosed myoepithelial carcinoma. Within 4 weeks postoperatively and 2 weeks into a chemotherapeutic regimen, the tumor exhibited progression. Radiation therapy was started and growth of the tumor was halted.

Myoepithelial carcinoma should be included in the differential diagnosis for clival masses, especially in patients with previously diagnosed myoepithelial carcinoma. The primary management of this tumor should be with chemotherapy and radiation, with surgery serving only for decompression.

Key Words • clivus • metastasis • myoepithelial carcinoma • transsphenoidal surgery • skull base • oncology
ing study demonstrated progression of the clival lesion (Fig. 4 left). At this point, she was started on CECT and underwent fractionated photon radiotherapy followed by proton beam radiotherapy to the clival mass over the course of 6 weeks, as suggested by preclinical data. Following 4 courses of CECT treatment over 4 months, on serial imaging the mass remained stable (Fig. 4 right), her sixth cranial nerve palsy was mostly resolved, and she remained otherwise neurologically stable. Unfortunately, she developed a malignant pleural effusion followed by a tension pneumothorax approximately 12 months after her clival decompression, and she died of complications of the metastatic myoepithelial carcinoma.

**Discussion**

The differential diagnosis of a clival mass is relatively broad, and includes metastatic lesions, lymphoma, chordoma, meningioma, pituitary adenoma, and nasopharyngeal carcinoma as well as a reconversion from yellow to red bone marrow. Our current case appears to be the first reported one of soft-tissue myoepithelial carcinoma metastatic to the clivus. In this case, suspicion was high for metastatic myoepithelial carcinoma in light of the patient’s prior history of metastasis to the lung. However, there have been reports of myoepithelial carcinoma that were metastatic at diagnosis, and such suspicion would not be present if the primary tumor was not identified prior to presentation with a clival mass. Therefore, we suggest that myoepithelial carcinoma of soft tissue could be considered in the differential diagnosis of a newly diagnosed clival mass.

Myoepithelial carcinoma of soft tissue is thought to be part of a continuum of tumor types that include mixed tumor, myoepithelioma, and parachordoma (and their malignant counterparts), which have been referred to as “MMP” tumors. Mixed tumors, or pleomorphic adenomas, are well-circumscribed lesions that contain both epithelial and myoepithelial elements. When malignant, these are referred to as malignant mixed tumors. Myoepitheliomas are composed primarily of myoepithelial cells that appear similar to those in mixed tumors; however, these tumors lack obvious ductal differentiation. When malignant, they are referred to as myoepithelial carcinomas. Parachordomas are similar in appearance to the mixed tumor but have the characteristic feature of prominent cytoplasmic vacuolization. Myoepithelial carcinomas are most often thought of as salivary gland tumors, having only recently been described as arising in the soft tissues. A recent cytogenetic analysis of 5 myoepitheliomas of soft tissue demonstrated rearrangement of the PLAG1 gene, which is the most common chromosomal aberration in mixed tumors of the salivary glands, and would point to a common pathway of development for salivary gland and soft-tissue myoepitheliomas. In a series of 101 patients with myoepithelial carcinoma of soft tissue, tumors were provisionally classified for the purposes of the study as either benign or malignant, based on the level of cellular atypia. Between those classified as benign, and those counted as malignant, there was a statistically significant difference in both recurrence rate and metastasis.

Myoepithelial carcinomas of soft tissue have not been reported to arise from preexisting soft-tissue malignancy; rather, they appear to arise de novo. These tumors may arise in many areas: commonly in the upper and lower limbs and limb girdles, head, neck, and torso, as well as the mediastinum, retroperitoneum, tricuspid valve, and lung. One case of myoepithelial carcinoma arising primarily in the sella turcica may be best classified as salivary in origin, given that pleomorphic adenoma arising from salivary gland rests has been reported in the posterior pituitary gland, and myoepithelial carcinoma from tumor would explain this intraxial primary site for this typically extraaxial malignancy.

Metastases from myoepithelial carcinomas of soft tissue have been reported to occur to the lung; mediasti-
num, lymph nodes, spine, orbit, soft tissue, bone, brain, bone marrow, and liver. Metastases at diagnosis are not uncommon and have been reported in the lungs, lymph nodes, and skin.2,5,7

Conclusions

This case represents a rare tumor metastasis that should be considered in the differential diagnosis of a clival mass when a history of myoepithelial carcinoma is present. This patient had a long and well-documented history of myoepithelial carcinoma, and therefore, preoperative suspicion was high that her clival mass represented metastasis of this malignancy. As mentioned previously, however, there is no reason to suggest that a clival metastasis would not present prior to an unnoticed primary lesion—which are typically painless masses or swelling—in other patients, and thus, myoepithelial carcinoma should be considered in the differential diagnosis of even a lone clival mass.

Once the diagnosis of myoepithelial carcinoma is made, decompression followed by medical and radiation management should be the standard, because this is an aggressive tumor that requires gross-total resection, which, especially in the situation of metastasis, is difficult.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Bohnstedt. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Bohnstedt. Study supervision: Shah.

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