Malignant Myoepithelioma in the Maxillary Sinus: Case Report and Review of the Literature

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Abstract. Malignant myoepithelioma of the head and neck usually arises in the salivary glands. We experienced a rare case with malignant myoepithelioma in the maxillary sinus. A 47-year-old woman with malignant myoepithelioma in the maxillary sinus underwent partial maxillectomy. However, local recurrence occurred 28 months after surgery and she was subsequently treated with radiation therapy with proton beams. The recurrent tumor showed complete response and the patient was alive with no evidence of disease 30 months after irradiation. No therapy-related severe toxicities were observed. A rare case with malignant myoepithelioma in the maxillary sinus was successfully treated with radiation therapy.

Malignant myoepitheliomas of the head and neck are known usually to arise in the major salivary glands, especially in the parotid glands, and less frequently, in the minor salivary glands (1, 2). Malignant myoepithelioma in the maxillary sinus is extremely rare. In our review of the literature, we discovered only three previously published cases (3-5).

We report the first case, to the best of our knowledge, of a patient with malignant myoepithelioma in the maxillary sinus who was successfully treated with radiation therapy.

Case Report

A 47-year-old woman had a 2-month history of progressive swelling and pain of the left cheek. Contrast-enhanced computed tomography (CT) revealed a homogenously enhanced mass in the left maxillary sinus. The mass was 40×30×30 mm in size and showed bone invasion into the medial and inferior walls of the left maxillary sinus. There was no cervical lymph node enlargement. Malignant cells were found in specimens obtained from a biopsy, and subsequently the patient underwent left partial maxillectomy. The tumor was completely resected macroscopically and was definitively diagnosed as malignant myoepithelioma by histopathological examinations including immunohistochemical staining: it was positive for S-100 protein, pankeratin, vimentin and glial fibrillary acidic protein (GFAP), while it was negative for smooth muscle actin (SMA) (Figure 1).

Twenty-eight months after surgery, local recurrence was detected by magnetic resonance imaging (MRI) and positron-emission tomography. The recurrent tumor was 40×40×40 mm and widely invaded into the left orbit, ethmoid sinus and subcutaneous tissues beyond the maxillary wall (Figure 2). Complete resection of the tumor was considered difficult and, furthermore, the patient refused a surgical procedure including enucleation of the eye. Consequently, radiation therapy was applied to the patient. Written informed consent was obtained from the patient before treatment.

Radiation therapy. The patient’s head was immobilized by a personally shaped head shell (ESFORM; Engineering System, Matsumoto, Japan). Treatment planning for radiation therapy was based on CT images at 3-mm intervals in the treatment position. The clinical target volume, which was...
defined as gross tumor volume plus a 5-mm margin, was homogeneously encompassed with 100% dose level. Proton beams of 150 MeV were used for treatment. In each treatment session, the positional relationship between the center of radiation fields and the adjacent structures was examined with the patient lying in the treatment position by the fluoroscopy unit attached to the treatment unit prior to irradiation. The beams were delivered through 3 ports using a rotational gantry (Figure 3). Fraction size was 2.2 GyE daily, 5 days per week, and the total dose was 79.2 GyE in 36 fractions. The overall treatment time was 56 days. With regard to therapy-related acute toxicities, only mild dermatitis and mucositis arose within the radiation fields through the treatment. These acute reactions were transient and easily manageable.

The irradiated tumor was gradually reduced, as shown by MRI at 4-month intervals, and disappeared 12 months after completion of irradiation (Figure 4). To date, no late toxicity associated with treatment has been observed. The visual acuity of bilateral eyes was normally preserved. The patient was alive with no evidence of disease at the last follow-up 30 months after radiation therapy.

Discussion

Approximately 10% of myoepitheliomas arise in the head and neck, especially in the salivary glands (1, 6). Myoepitheliomas account for 1% of all primary tumors in the salivary glands, and nearly 70% of them arise in the parotid gland, 20% in the submandibular gland, and 10% in the minor salivary glands (7, 8). Approximately 90% of myoepitheliomas are benign (1, 7). The pharynx, palate and tongue have been described in previously published case reports as rare primary sites of malignant myoepithelioma in the head and neck (9-11). These were considered as tumors of accessory salivary tissue origin. To the best of our knowledge, only 3 cases with malignant myoepithelioma in the maxillary sinus have been reported in the literature to date (Table I) (3-5). The patients were 60- and 67-year-old men and a 65-year-old woman. Of these patients, one underwent partial tumor resection, but died of intracranial invasion of the residual tumor 5 months after surgery. The remaining two patients were treated with radical maxillectomy. Of these two patients, one was alive with no evidence of disease 2 years after surgery, and the postoperative clinical course of the other patient is unknown.

Microscopically, myoepithelioma cells can be classically divided into 4 subtypes of epithelioid cells, spindle-shaped cells, plasmacytoid (hyaline) cells, and clear cells which the present tumor showed, based on morphology. Myoepithelioma is frequently lobulated and most commonly shows a reticular or trabecular structure with chondromyxoid or hyalinized stroma, and usually has no ductal lumen. Concerning immunohistochemical staining, myoepithelioma is generally positive for myoepithelial markers such as S-100 protein, pankeratin and vimentin, while being variable for SMA and GFAP. Savera et al. reported that the immunoreactive rates to myoepitheliomas of the salivary glands were 100% for S-100 protein, pankeratin and vimentin each, 50% for SMA and 31% for GFAP, respectively (12). The criteria to suggest malignancy of myoepithelioma include cytological atypia, cellular pleomorphism, mitotic activity, necrosis, and infiltrating growth. The present case was compatible with these histopathological findings and was finally diagnosed as malignant myoepithelioma.

Surgical resection is considered the most standard and curative modality for the treatment of malignant myoepithelioma in the major salivary glands (7, 8). To the best of our knowledge, there are no previous reports on treatment efficacies of radiation therapy alone for malignant myoepithelioma.

<table>
<thead>
<tr>
<th>Series</th>
<th>Age (years)</th>
<th>Gender</th>
<th>Tumor size (mm)</th>
<th>Treatment</th>
<th>Follow-up period (months)</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hsiao et al. 1997 (3)</td>
<td>60</td>
<td>M</td>
<td>90x70x35</td>
<td>Partial tumor resection</td>
<td>5</td>
<td>Dead of disease</td>
</tr>
<tr>
<td>Graadt van Roggen et al. 1998 (4)</td>
<td>67</td>
<td>M</td>
<td>60x50x40</td>
<td>Radical maxillectomy</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Sunami et al. 1999 (5)</td>
<td>65</td>
<td>F</td>
<td>70x70x70</td>
<td>Radical maxillectomy</td>
<td>24</td>
<td>Alive with NED</td>
</tr>
<tr>
<td>Present case</td>
<td>47</td>
<td>F</td>
<td>40x30x30</td>
<td>Partial maxillectomy</td>
<td>28</td>
<td>Recurrence</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>40x40x40</td>
<td>Radiation therapy for local recurrence</td>
<td>30</td>
<td>Alive with NED</td>
</tr>
</tbody>
</table>

M, Male; F, female; NA, not available; NED, no evidence of disease.
Figure 1. Photomicrographs of specimens of the tumor. a, The tumor shows proliferative invasion into the normal tissues along with lobular and trabecular structures (hematoxylin-eosin stain; original magnification, ×100). b, The tumor cells have a clear cytoplasm and irregular nucleus, and hyalinized stroma lies between them. There is no luminal cell component, and a few mitoses are observed (hematoxylin-eosin stain; original magnification, ×400). c, The tumor cells, which strongly stain brown, are immunoreactive for S-100 protein (S-100 protein stain; original magnification, ×100). d, Parts of the tumor cells, which slightly stain brown, are immunoreactive for glial fibrillary acidic protein (GFAP) (GFAP stain; original magnification, ×100).
myoepithelioma. Radiation therapy with proton beams was applied to the present case with a curative intent. Proton beam irradiation can theoretically produce excellent dose localization to the target compared with photon (X-ray) irradiation, due to the sharp distal fall-off of the Bragg peak of proton beams (13, 14). It decreases the irradiated volume and doses given to the adjacent critical organs, while increasing the dose to the target. Therefore, it is expected to result in enhanced efficacy and lower toxicity. Indeed, in the present case, a high dose of 79.2 GyE was safely delivered to the tumor adjacent to the brain and contra-lateral eye. Radiation therapy appeared effective for local control in this patient although the follow-up period of 30 months was relatively short.

Conclusion

We experienced a rare case with malignant myoepithelioma in the maxillary sinus, which was successfully treated with radiation therapy. Radiation therapy may be a useful treatment modality for malignant myoepithelioma, especially unresectable tumors, or for patients who are medically inoperable or refuse surgical resection. Further studies are required to define the role of radiation therapy for malignant myoepithelioma.

Acknowledgements

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Figure 4. Coronal T2-weighted MR image 12 months after irradiation. The tumor has completely disappeared, and the left eye ball is restored to its normal position.

References