Hidradenocarcinoma on the Orbit

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INTRODUCTION

Hidradenocarcinoma is a malignant tumor that originates from the eccrine gland, and it has a very aggressive pattern. Local invasion and distant metastasis occur frequently, and the recurrence rate after treatment is also high¹,₂,⁶. So far, no standard treatment has been established because the incidence of hidradenocarcinoma is very low. The treatment option which most researchers have recommended is a complete resection²,¹³. Adjuvant treatment including radiotherapy and chemotherapy also can be tried, but the effectiveness of these treatment regimens is not clear.

CASE REPORT

A 40-year-old man presented with exophthalmus for 8 months and decreased vision for 1 month on left eye. The ophthalmological examination of left eye revealed visual field defect and decreased vision (0.2) and increased intraocular pressure (20 mmHg). Computed tomography (CT) showed a well enhancing tumor in the left orbit with focal bone destruction of lateral orbital wall and focal invasion into the anterior portion of left cavernous sinus. Magnetic resonance image (MRI) revealed a localized mass at lateral aspect of left orbit measuring 4.5×2.2×3.6 cm³ in size which is iso-signal intensity on T1 and T2-weighted image, and heterogeneous enhancement at lateral aspect of left orbit. The focal bone destruction was seen at lateral orbital wall. The mass invaded into temporalis muscle, superior orbital fissure, and cavernous sinus (Fig. 1A-C). We performed a biopsy and the hidradenocarcinoma was diagnosed; hence, we planned to surgical resection with enucleation of eye ball. The grossly total resection was done through the modified lateral supraorbital approach (Fig. 2). The tumor appeared hyperemic and yellowish mass. Dissection the optic nerve

Fig. 1. Preoperative and postoperative magnetic resonance imaging (MRI) findings. (A-C) Preoperative MRI and computed tomography showed a heterogeneous enhanced mass with focal bony destruction. (D, E) The immediate postoperative MRI showed no residual tumor.
from the tumor was difficult. The left eye ball was enucleated
and fat packing was done. The immediate postoperative MRI
showed no residual tumor.

In the biopsy of the tissue removed by surgery, ill-defined

multilobular tumors were extensively observed in dermis. The
tumors were surrounded by pseudocapsules consisting with
fibrotic stroma, and a cystic cavity caused by tumor necrosis
was also observed. The tumor cells were round to polygonal
in shape with a clear cytoplasm. Cellular atypia and mitosis
were observed frequently. Additionally, we performed immuno-
histochemical staining, and the tumors were positive for epil-
theelial membrane antigen (EMA), vimentin, cytokeratin, S-100,
and high proliferation labeling index (Ki-67 30%), but they were
negative for carcinoembryonic antigen (CEA) and gross cystic
disease fluid protein 15 (GCDFP-15) (Fig. 3). Based on these
histological findings and imaging study, we had diagnosed as
hidradenocarcinoma on left upper eyelid extending into the
left cavernous sinus with orbital bony destruction.

Considering the characteristics such as a very high recurrence
rate and well metastasis of the tumor, adjuvant radiotherapy
was performed with a total 5,400 cGy/27 fraction. The patient’s
status had been well maintained for 4 months without recur-
rence and complications.

The patient presented the sudden right eye visual symptom
at 5 months after surgery. The ophthalmological examination
of right eye revealed visual field defect and decreased vision
(only light perception) and 16 mmHg intraocular pressure. The
mydriatic test result found a focal atrophy in the retina. In
addition, the visual evoked potential result was that the P1
and P3 latency was delayed, and the electoretinography result
found that the rod function was decreased. However, the multi-
focal electoretinogram was normal and both the MRI and
positron emission tomography (PET-CT) image showed no
recurrence and no metastasis (Fig. 4). We considered that the
visual problem was associated with cancer associated retino-
pathy (CAR). And we started systemic steroid therapy and ad-
juvant chemotherapy. We administrated cisplatin and 5-fluorou-
racil. After the 1st cycle of chemotherapy, the patient’s cognitive
function was getting worse and other cranial nerve (III, IV, VI)
palsy was developed. We recommended the MRI evaluation
and 2nd cycle of chemotherapy, but the patient refused the
additional therapy. The patient expired at 7 month later.

DISCUSSION

Hidradenocarcinoma was first described by Keasbey and
Hadley in 1954. It is a very rare tumor, and Keasbey found
only 3 cases of hidradenocarcinoma among the 235 sweat
gland tumors when he first described this tumor in 1954.
Mehregan et al.,[1] mentioned that only 2 cases were diagnosed
as hidradenocarcinoma among the 450,000 tissue samples
collected for 20 years in the dermatopathology laboratory, and
its prevalence is less than 0.001%.1[1] So far, less than 70 cases
have been reported. Patients with hidradenocarcinoma present
with non-specific clinical symptoms. In most cases, a solitary
cutaneous or subcutaneous lump for a long time period and
the tumor size increases gradually. The tumor is often accom-
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panied by ulcers, and occasionally there are a variety of clinical manifestations such as fleshly color or red to gray color. It may occur anywhere including the head, torso, legs, and arms; but, it mainly occurs on the head and neck according to the reports. Also most of the tumor shows rapid growth over weeks to months, and it can cause aggressive local invasion and metastasis to the surrounding lymph nodes or other distant organs. In fact, hidradenocarcinoma has a poor prognosis, and the disease-free survival rate within 5 years is less than 30% according to the literature, and most of the reports suggested that distant metastasis is observed after tumor recurrence and eventually it leads to death. Statistically, the local recurrence rate after surgical resection is approximately 50%, and more than 60% of the patients show metastasis to the lymph nodes, bones, skin, and internal organs.

Hidradenocarcinoma is a histopathologically ill-defined multilobular solid tumor located from the dermis to the subcutaneous tissue, and tumor necrosis is often observed in the lobule. The polygonal-shaped cells have oval nuclei and clear to slightly eosinophilic cytoplasm. The tumor cells show significant nuclear atypia and moderate mitotic activity. According to the literature, the immunohistochemical findings of hidradenocarcinoma are different, but most of the tumors are positive for vimentin, EMA, cytokeratin 5/6 (CK5/6), and they show various responses to S-100, Ki-67, and GCDFP-15. In this case, the tumor was microscopically diagnosed as hidradenocarcinoma which was positive for vimentin, EMA, CK5/6, S-100, and high proliferation labeling index (Ki-67 30%) and negative for GCDFP-15 and CEA.

No standard treatment has been established so far because hidradenocarcinoma has a very low incidence, and it is also challenging to treat the tumor due to its aggressive clinical characteristics. Wide resection is performed in most of the cases, and adjuvant therapy is considered as an additional treatment option. However, the prognosis of patients and clinical features still remain a matter of debate. The wide local excision method may be a suitable local treatment option for non-disseminated lesions. However, although there is no evidence of distant metastasis and invasion of local lymph nodes on PET-CT, there is a possibility of microscopic invasion of lymph nodes in cases of locally aggressive invasion as in this case. Also, considering the high recurrence and metastatic rates of hidradenocarcinoma after surgery, we considered that combined treatment with wide excision and adjuvant radiotherapy would be appropriate for this patient. We performed adjuvant radiotherapy after complete surgical resection, and the patient is doing well and there has been no recurrence and complications for 4 months after surgery. However, 5 months after surgery, the patient’s opposite eye showed symptoms of CAR. We administered chemotherapy, but 7 months later the patient was dead, CAR is a rare phenomenon associated with existing malignant tumor, and there is no standard treatment guideline. Appearance of CAR can imply that there can be a remaining malignant cells. In the case CAR occurring in the patient’s opposite eye, we can consider that it would be a systemic metastasis.

We performed a gross total resection with postoperative radiotherapy and chemotherapy. And the patient was doing well for a relatively long time, considering the aggressive progression of hidradenocarcinoma. But, the disease progression was rapidly worsen after appearance of CAR. So, we considered that when hidradenocarcinoma is combined with cancer associated functional loss, it can act a poor prognosis factor.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES


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