Malignant Fibrous Histiocytoma of the Lateral Wall of the Orbit

—Case Report—

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Abstract

A 58-year-old male presented with malignant fibrous histiocytoma originating from the lateral wall of the orbit, manifesting as left exophthalmos and diplopia on left lateral gaze. Magnetic resonance (MR) imaging demonstrated the tumor extending into the left orbit and under the temporalis muscle. The tumor was isointense with muscle on T₁-weighted MR images and was surrounded by a low-intensity rim, which was proved to be the capsule after histological examination. The tumor was completely removed after detaching the lateral orbital rim and zygomatic arch using the dismasking flap method. He underwent one course of CYVADIC chemotherapy and has demonstrated no recurrence for 1 year and 6 months postoperatively.

Key words: malignant fibrous histiocytoma, dismasking flap, lateral wall of the orbit

Introduction

The concept of malignant fibrous histiocytoma (MFH) was introduced in the early 1960s to describe a group of soft tissue tumors which were supposed to be derived from histiocytes.^{10,12,16)} MFH accounts for 10.5% of all soft tissue sarcomas, and about 0.1% of all malignant tumors.^{14,17)} MFH consists of fibroblast-like and histiocyte-like cells, although the relative populations and organization of these cells vary in each tumor. However, a recent immunohistochemical study demonstrated that MFH is independent of macrophages or monocytes derived from histiocytes and suggested that fibroblasts are the tumor origin.²²⁾ Therefore, the histogenesis of MFH is currently confused and controversial. Enzinger and Weiss²⁾ proposed a widely used classification in 1988 which divides MFHs into five types: storiform-pleomorphic, myxoid, giant cell, inflammatory, and angiomatoid. The storiform-

pleomorphic type is the most common, accounting for over two-thirds of all MFHs.¹⁹⁾ MFH can arise from the soft tissues supporting the structures of many organs and tissues. MFH occurs in the extremities in 68% of cases, the retroperitoneal and abdominal areas in 16%, and the trunk in 9%, but is relatively rare in the head and neck region.²⁰⁾ MFH in the head and neck region occurs in the sinonasal tract in 30% of cases, the craniofacial bones in 15–20%, the larynx in 10–15%, the soft tissue of the neck in 10–15%, the major salivary glands in 5–15%, and the oral cavity in 5–15%.¹⁾ Solitary cases of MFH have arisen from the mandible,¹¹⁾ the eyelid,¹³⁾ and the brain.

We present a case of MFH originating from the lateral wall of the orbit, extending into the orbit and under the temporalis muscle.

Case Report

A 58-year-old male presented with a 2-month history of diplopia and recent exacerbation of left exophthalmos. On admission to Matsue Municipal

Fig. 1 Precontrast axial CT scan showing a mass extending into the left orbit and under the temporalis muscle. The left lateral wall of the orbit and major sphenoidal wing are partially destroyed and the left eyeball is pushed forward.

Fig. 2 Coronal T_1 -weighted MR images showing the mass with a low-intensity rim. The tumor intensity is similar to that of muscle (left). The tumor is enhanced by Gd-DTPA administration except for the central area (right).

Hospital on July 4, 1994, left exophthalmos and swelling of the left cheek were observed. Abduction of the left eyeball was limited on lateral gaze. Visual acuity was 3/10 in the right eye and 2/10 in the left. Hertel exophthalmometry recorded 18 mm on the right and 22.5 mm on the left. Other physical and neurological examinations were normal. Laboratory studies did not detect any abnormalities.

Computed tomography (CT) showed a bony defect measuring 1.6×2.1 cm in the left lateral wall

Fig. 3 Schema for the surgical procedure. Solid lines indicate the skin incision, broken lines show the osteotomies, and the dotted line indicates the midline.

of the orbit, and an isodense mass measuring $2.4 \times 1.4 \times 2.3$ cm in the orbit and $2.7 \times 1.8 \times 7$ cm under the temporalis muscle (Fig. 1). T_1 -weighted magnetic resonance (MR) imaging revealed a well-defined tumor with a low-intensity rim. The tumor was isointense with the temporalis muscle and was homogeneously enhanced after gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA) administration, but the central area of the tumor remained unenhanced (Fig. 2). Left external carotid angiography demonstrated that the lesion was stained slightly via a left deep temporal artery. Scintigraphy using technetium-99m-methylene diphosphonate ($^{99m}\text{Tc-MDP}$) and gallium-67 showed no uptake other than that in the lesion. An aspiration biopsy was performed under local anesthesia. The histological diagnosis was a soft tissue sarcoma, since the specimen consisted of atypical spindle cells, *i.e.* fibroblast-like cells.

A semi-coronal skin incision was made, starting 1 cm below the left zygomatic arch and extending to the midline of forehead. A circum-palpebral blepharoplasty incision was added and dissection was performed up to the orbital rim, which allowed the left orbital structures including the left eyeball to be detached from the skin flap (dismasking flap method,¹⁸⁾ Fig. 3). The whitish-gray tumor appeared after the skin flap and the temporalis muscle were reflected downward. The tumor was easily isolated from the muscle, possibly because of the presence of a capsule, which appeared as the low-intensity rim

Fig. 4 Photomicrograph showing cellular connective tissue with fibroblastic spindle-shaped cells arranged in a storiform pattern. HE stain, $\times 200$.

on MR images. The frontotemporal craniotomy was followed by detaching the lateral part of the orbital rim and the anterior part of the zygomatic arch using three osteotomies (Fig. 3). The tumor was removed en bloc because it could be easily isolated from the periorbital and surrounding structures. The lateral wall of the orbit was reconstructed with the inner plate of the frontotemporal bone. The three pieces of bone were fixed using titanium microplates. The supraorbital and supratrochlear nerves, which were cut at the stage of skin flap reflection, were anastomosed.

Histological examination of the specimens showed fibroblastic spindle-shaped cells arranged in a storiform pattern and histiocyte-like cells among them (Fig. 4). The peripheral part of the tumor contained fibrous capsule-like structures, but no infiltrated malignant cells. No other type of soft tissue differentiation such as osteocyte, chondrocyte, or leiomyocyte was observed. Immunohistochemical study found some tumor cells were positive for α -1-antichymotrypsin and α -smooth muscle actin. No tumor cells were immunoreactive for cytokeratin, epithelial membrane antigen, vimentin, desmin, lysozyme, α -1-antitrypsin, or S-100 protein. The tumor was diagnosed as the storiform-pleomorphic type of MFH. Chromosomal analysis of the MFH cells revealed 51 chromosomes including X and Y. Abnormal features were ?add(1)(p36), -5, -7, +19, +6 marker chromosomes (Fig. 5).

Postoperatively, the left exophthalmos, swelling of the left cheek, and diplopia on left lateral gaze disappeared. Numbness of the forehead due to cutting the supraorbital nerve disappeared 2 months after surgery. He received CYVADIC chemotherapy³⁾ us-

Fig. 5 Photomicrograph showing chromosomal analysis of the tumor cell, revealing 51, XY, ?add(1)(p36), -5, -7, +19, +6 marker chromosomes.

Fig. 6 Coronal T_1 -weighted MR image with Gd-DTPA obtained 1 year after surgery. The intraorbital structures have returned to their original positions and no tumor recurrence is observed. Similar findings were seen 1 year and 6 months after surgery.

ing total doses of 800 mg of cyclophosphamide, 2.5 mg of vincristine, 70 mg of adriamycin, and 2000 mg of dactinomycin administered intravenously over 5 days. He has not demonstrated recurrence for 1 year and 6 months (Fig. 6).

Discussion

This is the first reported case of MFH originating from the lateral wall of the orbit. The most probable origin of the tumor was the bone of the lateral wall of the orbit, but other tissue such as the temporalis muscle could not be excluded, since the MFH could have originated from any soft tissue around the lateral wall of the orbit. The tumor had formed a pseudo-membrane and therefore seemed to be isolated from the surrounding structures. The patient presented with swelling of the left cheek, left exophthalmos, and diplopia on left lateral gaze due to the presence of the tumor mass. Pain was absent, which is typical of MFH. These signs and symptoms disappeared after total removal of the tumor. The tumor was isodense with muscle on CT scans and was enhanced slightly by contrast administration. CT was especially useful for detection of the bony defect. MR imaging showed that the tumor was isointense with muscle on the T₁-weighted image with enhancement, and hyperintense on the T₂-weighted image, as reported in other cases.⁷⁾ The cranial tumor was assumed to be the primary lesion, because ^{99m}Tc-MDP and gallium-67 uptake was limited to the lesion. Histological examination demonstrated a typical storiform MFH with a smaller pleomorphic region. Immunohistochemical examination showed positive staining for α -1-antichymotrypsin and α -smooth muscle actin. Meister⁹⁾ reported that α -1-antichymotrypsin was more specific as a tumor marker for MFH than lysozyme, although no other markers were pathognomonic for MFH. Chromosomal analysis found only one ring chromosome among the common features in histiocytomas such as rings, dicentrics, and telomeric association.⁸⁾

Weiss and Enzinger²¹⁾ reported that MFH is highly malignant, with 44% incidences for both recurrence and metastasis, and the survival rate at 2 years was 60%. Kearny *et al.*⁵⁾ found local recurrence in 51% of patients despite total removal of the tumor, but radical surgery improved survival to 68% at 5 years.¹⁵⁾ Recurrence and metastasis rates were 44% and 42% in 200 cases of the storiform-pleomorphic type, 66% and 23% in the 80 myxoid type cases, both less than 50% in the 32 giant cell type cases, and approximately 50% and 33% in the inflammatory type cases, respectively.²¹⁾ However, Limacher *et al.*⁶⁾ reported that the myxoid and angiomatoid types had a better prognosis. Histological features of malignancy such as bizarre giant cells, anaplasia, number of mitotic figures, and necrosis have not been verified as predictors. The size of the tumor and its depth are also factors affecting the outcome.⁵⁾ Hashimoto⁴⁾ reported

that the 5-year survival rate in patients with predominantly storiform pattern was 70.8%, compared to 47.7% for all MFHs. MFH tends to form a pseudo-membrane but infiltrates adjacent tissues across the membrane, so radical and wide removal of the MFH is mandatory. Radiation and chemotherapy are not the first choice of treatment for patients with MFH, but may be useful for patients with poor surgical risk or more than two lesions, and may also be used in addition to surgery.

We enucleated the tumor as generally used in cases of MFH, although the tumor was diagnosed as MFH before surgery. The dismasking flap method was used to detach the orbital structures from skin flap. This procedure exposed a sufficiently wide region of the orbital rim and zygomatic arch without hard retraction of the skin, which is harmful to the eyeball, optic nerve, facial nerve, jaw joint, *etc.*, resulting in no cosmetic or functional problems. After adjuvant CYVADIC chemotherapy, no local recurrence or metastasis to the local lymph nodes or lung has been observed.

References

- 1) Barnes L, Kanbour A: Malignant fibrous histiocytomas of the head and neck. A report of 12 cases. *Arch Otolaryngol Head Neck Surg* 114: 1149-1156, 1988
- 2) Enzinger FM, Weiss SW: *Soft Tissue Tumors*, ed 2. St Louis, CV Mosby, 1988, pp 269-300
- 3) Gottlieb JA, Baker LH, O'Bryan RM, Sinkovics JG, Hoogstraten B, Quagliana JM, Rivkin SE, Bodey GP, Rodriguez VT, Blumenschein GR, Saiki JH, Coltman C, Burgess MA, Sullivan P, Thigpen T, Bottemley R, Balcerzak S, Moon TE: Adriamycin used alone and in combination for soft tissue and bone sarcomas. *Cancer Chemotherapy Reports* 6: 271-282, 1975
- 4) Hashimoto H: Malignant fibrous histiocytoma. A clinicopathologic study of 130 cases. *Fukuoka Igaku Zasshi* 70: 585-613, 1979 (in Japanese)
- 5) Kearny MM, Soule EH, Ivins JC: Malignant fibrous histiocytoma. A retrospective study of 167 cases. *Cancer* 45: 167-178, 1980
- 6) Limacher J, Delage C, Lagace R: Malignant fibrous histiocytoma. Clinicopathologic and ultrastructural study of 12 cases. *Am J Surg Pathol* 2: 265-274, 1978
- 7) Mahajan H, Kim EE, Wallace S, Abello R, Benjamin R, Evans HL: Magnetic resonance image of malignant fibrous histiocytoma. *Magn Reson Imaging* 7: 283-288, 1989
- 8) Mandahl N, Heim S, Arheden K, Rydholm A, Willen H, Mitelman F: Rings, dicentrics, and telomeric association in histiocytomas. *Cancer Genet Cytogenet* 30: 23-33, 1988
- 9) Meister P: Malignant fibrous histiocytoma. History,

- histology, histogenesis. *Pathol Res Pract* 183: 1-7, 1988
- 10) O'Brien JE, Stout AP: Malignant fibrous xanthomas. *Cancer* 17: 1445-1458, 1964
 - 11) Onyango JF, Awange DO: Primary malignant fibrous histiocytoma of the mandible. A case report. *East Afr Med J* 70: 460-463, 1993
 - 12) Ozzelo L, Stout AP, Murray MR: Cultural characteristics of malignant histiocytomas and fibrous xanthomas. *Cancer* 16: 331-344, 1963
 - 13) Roth AM: Malignant fibrous histiocytoma of the ocular adnexa. A case report. *Metab Pediatr Syst Ophthalmol* 16: 5-8, 1993
 - 14) Russel WO, Cohen J, Enzinger FM, Hajdu SI, Heise H, Harting RC, Meissner W, Miller WD, Schmitz RL, Suite HD: A clinical and pathological staging system for soft tissue sarcomas. *Cancer* 40: 1562-1570, 1977
 - 15) Soule EH, Enriquez P: Atypical fibrous histiocytoma, malignant fibrous histiocytoma, malignant histiocytoma, and epithelioid sarcoma. A comparative study of 65 tumors. *Cancer* 30: 128-143, 1972
 - 16) Stout AP, Lattes R: Tumors of the soft tissues, in: *Atlas of Tumor Pathology, ser 2, fasc 1*. Washington, DC, Armed Forces Institute of Pathology, 1967, pp 38-52, 107-115
 - 17) Suit HD: Sarcoma of soft tissue. Clinical and histopathological parameters and response to treatment. *Cancer* 35: 1478-1483, 1975
 - 18) Tajima S, Tanaka Y, Imai K, Ueda K, Maejima S, Yabu K: Extended coronal flap-dismasking flap for craniofacial and skull base surgery. *Bull Osaka Med Coll* 39: 1-8, 1993
 - 19) Weiss SW: Malignant fibrous histiocytoma. A reaffirmation. *Am J Surg Pathol* 6: 773-784, 1982
 - 20) Weiss SW, Enzinger FM: Myxoid variation of malignant fibrous histiocytoma. *Cancer* 39: 1672-1685, 1977
 - 21) Weiss SW, Enzinger FM: Malignant fibrous histiocytoma. An analysis of 200 cases. *Cancer* 41: 2250-2266, 1978
 - 22) Wood GS, Bekstead JH, Turner RR, Hendrickson MR, Kempson RL, Warnke RA: Malignant fibrous histiocytoma tumor cells resemble fibroblasts. *Am J Surg Pathol* 10: 323-325, 1986

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