Orbital Malignant Fibrous Histiocytoma With Extension to the Base of the Skull

—Case Report—

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Abstract

An 18-year-old woman presented with a malignant fibrous histiocytoma (MFH) originating in the orbit and invading the frontal and temporal base of the skull manifesting as exophthalmos and double vision that had persisted for 2 months. Magnetic resonance imaging revealed a tumor in the left orbit that extended as far as the frontal and temporal base of the skull. The tumor was treated by radical resection with conservation of the eyeball and its contents, followed by orbit wall reconstruction. The histological diagnosis was MFH. Local radiotherapy was administered postoperatively. The preoperative symptoms improved, and there has been no evidence of local recurrence or metastasis in the year since the surgery. In this case, radical resection of the tumor was essential. Furthermore, the adjuvant therapy was apparently successful, probably because this histological type of tumor is highly sensitive to radiotherapy.

Key words: orbital tumor, malignant fibrous histiocytoma, sphenoid bone

Introduction

Malignant fibrous histiocytoma (MFH) is the most common of the malignant tumors of soft tissue, consisting of fibroblast-like and histiocyte-like cells.11) MFH of bone is also known.4) The origin of this type of histiocytoma is unclear, but the dominant hypothesis is undifferentiated mesenchymal cells.14) The highest incidence is in males between 50 and 70 years of age. The most common sites of MFH are the extremities (68%) and the retroperitoneal area (16%), and very rarely in the head and neck region (9%).2,14) Only 23 cases of MFH in the orbit have been reported.1,2,5,6,9,10,12) We encountered a case of MFH that arose in the orbit and invaded the frontal and temporal base of the skull.

Case Report

An 18-year-old woman first noted exophthalmos in her left eye after it was pointed out to her by a friend in early November 2000. The exophthalmos rapidly became more prominent and she developed progressive double vision, so she consulted a local doctor on November 21. Magnetic resonance (MR) imaging revealed a tumor in the left orbit, and she was referred to our hospital. Her past history and family history were unremarkable.

Physical examination on admission revealed exophthalmos (20 mm on the left and 12 mm on the right by Hertel exophthalmometry). Neurological examination, including examination of visual acuity, visual field examination by Goldmann perimeter, and funduscopy, yielded no abnormal findings, but the patient experienced double vision upon gazing upward. Biochemical tests of the blood, including measurement of tumor markers, showed no abnormal findings. Computed tomography (CT) with contrast medium of the thorax and abdomen revealed no abnormal findings. Whole-body scintigraphy with gallium-67 and bone scintigraphy with technetium-99m-methylene diphosphonate showed no abnormal intake except in the left periorbital region.

Preoperative CT revealed a 4.5 cm × 2.5 cm tumor of uniform density, except for one low-density area, located in the superolateral portion of the left orbit. CT also revealed bone destruction by the tumor in the posterior wall, lateral wall, and roof...
Fig. 1  T1-weighted magnetic resonance images with gadolinium-diethylenetriaminepenta-acetic acid revealing a tumor with heterogeneous enhancement, located in the superolateral portion of the orbit and the extramuscle cone, compressing the eyeball anteriorly and the optic nerve and extraocular muscles medially, and invading the frontal and temporal base of the skull (A, axial section; B, sagittal section).

Fig. 2  Selective left external carotid angiogram showing a hypervascular tumor fed by the left middle meningeal artery.

of the left orbit. Preoperative T1-weighted MR imaging with gadolinium-diethylenetriaminepenta-acetic acid showed a tumor located in the superolateral portion of the left orbit and the extramuscle cone, which compressed the left eyeball anteriorly and the optic nerve and the extraocular muscles medially, and invaded the frontal and temporal base of the skull with inhomogeneous enhancement (Fig. 1). Selective left external carotid angiography showed a highly vascularized tumor fed by the left middle meningeal artery (Fig. 2). The preoperative diagnosis was osteosarcoma or chondrosarcoma.

A left frontotemporal craniotomy was performed on December 12, 2000. The frontotemporal craniotomy revealed a grayish-red, easily bleeding tumor with an elastic consistency that had invaded the frontal and temporal base of the skull. The tumor had destroyed the lateral wall of the orbit and was located in the region immediately inferior to the temporal muscle. Histological examination of a biopsy specimen demonstrated MFH. The tumor was located outside the periorbita, so was surgically resected while conserving the orbital contents, including the periorbita. The roof and posterior wall of the orbit, which appeared to be the source of the tumor, were also resected, and the dura at the temporal tip where tumor infiltration was detected was also resected. As a result, the tumor was totally removed macroscopically. The orbit was reconstructed with artificial bone.

Histological examination showed markedly pleomorphic and heteromorphic cells that had densely proliferated (Fig. 3A, B). Mitotic figures were noted in the cells. Immunohistochemical studies revealed that the tumor cells were positive for CD68 (Fig. 3C) and α1-antitrypsin (Fig. 3D), and negative for S-100 protein, desmin, and muscle actin. The diagnosis was the storiform-pleomorphic type of MFH.

Postoperatively, 60 Gy local radiotherapy was administered to the region from which the tumor had been removed. The exophthalmos and double vision improved. MR imaging of the skull and total-body CT performed 1 year postoperatively showed no evidence of tumor recurrence or metastasis (Fig. 4).

Discussion

Although we are uncertain of the origin of the MFH tumor in the present case, the findings support two possibilities. The bone destruction and manner of tumor progression suggested that this tumor most likely originated in the wall of the orbit. However, the intraoperative findings could not exclude the soft tissue peripheral to the orbital walls as another possible source of the tumor. Only one case of MFH that originated in the orbital wall has been reported, so this is a very rare tumor.

MFH can be classified into four types based on a variety of histological features: storiform-pleomorphic, myxoid, giant cell, and inflammatory. The efficacy of radiotherapy is related to the histological features of the tumor, and may be more effec-
Fig. 3 Photomicrographs of the tumor specimen. A: The spindle-shaped cells were randomly arranged and showed marked pleomorphism and bizarre nuclei. Hematoxylin and eosin stain, ×100. B: The tumor consisted of a mixture of spindle-shaped and round histiocyte-like cells with bizarre chromatin patterns. Hematoxylin and eosin stain, ×400. C: Immunohistochemical examination showed the large tumor cells expressed CD68 especially strongly. ×400. D: Immunohistochemical examination showed most tumor cells expressed α1-antitrypsin. ×400.

Fig. 4 T1-weighted magnetic resonance images with gadolinium-diethylenetriaminepenta-acetic acid performed 1 year postoperatively showing improvement of the exophthalmos and no evidence of tumor recurrence (A, axial section; B, coronal section).

Radical tumor resection that includes the surrounding area is essential for the treatment of the tumor.8) In the
present case, radical resection of the tumor was performed and radiotherapy was administered postoperatively. The patient had no evidence of recurrence or metastasis at one year following surgery. However, since the prognosis for patients with this tumor is very poor, the patient must be followed up with careful monitoring for possible changes at the site of resection and throughout the body.

Although wide resection is the treatment of choice, tumors of the head, in contrast to tumors in other regions, pose functional problems that often make wide resection difficult. Therefore, postoperative adjuvant therapy is very important in the treatment of tumors of the head. Better understanding of the relationship between the various histological features of MFH and the efficacy of radiotherapy and/or chemotherapy would make adjuvant therapy more useful, thus achieving better outcomes without residual cosmetic and functional problems.

References


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