

## —Report on Experiments and Clinical Cases—

## Malignant Fibrous Histiocytoma of the Hypopharynx: A Case Report in a Young Adult

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### Abstract

This is the first case report of malignant fibrous histiocytoma of the hypopharynx in a young adult with preservation of the larynx. Although the initial histopathological diagnosis of the biopsy was benign fibrous histiocytoma, subsequent histopathological evaluation of the surgical specimen was malignant fibrous histiocytoma. Thereafter, re-excision of the histiocytoma with a wide margin was performed coupled with reconstruction of the hypopharynx while preserving the larynx. We also report the good long-term survival of this case of low-grade type of malignant fibrous histiocytoma.

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**Key words:** malignant fibrous histiocytoma, low-grade malignancy, hypopharynx, young adult, partial hypopharyngectomy

### Introduction

Malignant fibrous histiocytoma (MFH) is the most common type of bony and soft tissue sarcoma in middle-aged or elderly adults<sup>1-3</sup>. However, it does not commonly occur in young adults especially in the head and neck region, notably, the pharynx. Until now, only three cases of advanced MFH have been reported in the hypopharynx and oropharynx in middle-aged and elderly adults<sup>4-6</sup>. This is the first report of a young adult with MFH in the hypopharynx who underwent surgery with preservation of the larynx.

### Case Report

A 29-year-old male was referred to the

Department of Otolaryngology of Nippon Medical School Hospital on September 30, 1997 with an 8-month history of discomfort in the throat. He also presented with symptoms of bloody sputum during the one month prior to his visit to our hospital but without any associated complaints of hoarseness, dysphagia or dyspnea. Fiberlaryngoscopy revealed a red submucosal mass in the left pyriform sinus of the hypopharynx. The base of the mass was formed by the posterior wall of the pyriform sinus without fixation of the left vocal cord. Esophagogram showed an oblong mass, 3.5 cm in size, in the left pyriform sinus (**Fig. 1**). CT scan revealed a 3.5 × 1.5 cm mass in the posterior and left lateral wall of the hypopharynx (**Fig. 2**). Two biopsy specimens of the hypopharynx were taken by direct esophagoscopy under general anesthesia. The tumor was hard at biopsy. Microscopically, the biopsy specimens

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Fig. 1 Esophagography

A round mass was observed in the left pyriform sinus in the hypopharynx. The size of the tumor was 3.5 cm at its largest diameter (**arrow head**).

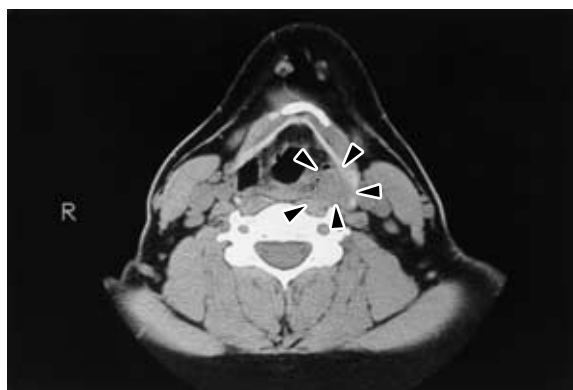


Fig. 2 CT scan of the hypopharynx

A round mass in the left pyriform sinus in the hypopharynx at the level of the greater ala of the thyroid cartilage was observed (**arrow head**). An air space can be seen between the anterior wall of the hypopharynx and the tumor.

showed abundant, but rather loose, spindle shaped cells arranged in fascicular and occasionally storiform patterns. Immuno-histologically, the tumor cells were strongly positive for vimentin, partially for CD-68 (KP-1) and faint for anti- $\alpha$ 1 tripsin. However, they were all negative for CD-34, desmin,  $\alpha$ -smooth muscle actin, and S-100 protein. No mitosis

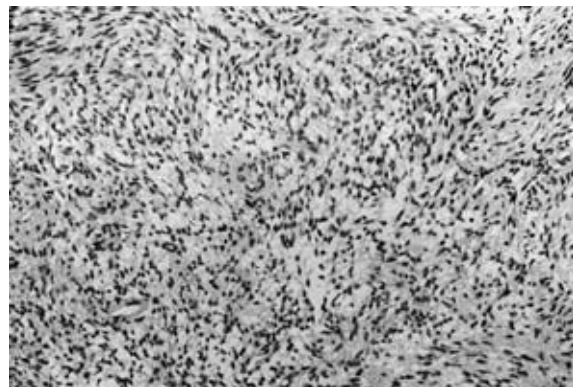


Fig. 3 Histological findings at the initial biopsy (HE  $\times$  20)

Loose proliferation of spindle shaped cells without mitosis is observed.

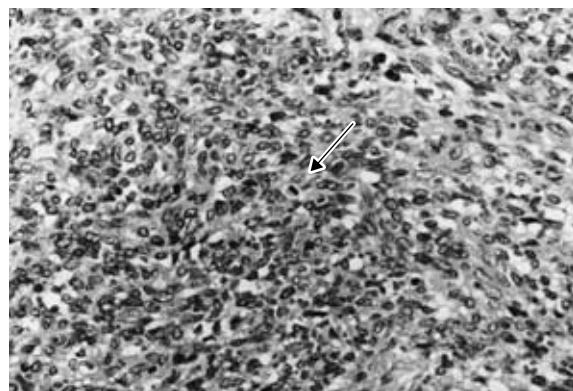


Fig. 4 Histological findings at the partial hypopharyngectomy (HE  $\times$  40)

Dense proliferation of spindle shaped cells and mitotic figure (**arrow**) are observed. The MIB-I index was 1~5% using Ki-67 (proliferation marker) by immuno-histological staining.

was observed. The histopathological diagnosis of the biopsy was benign fibrous histiocytoma (BFH) of the hypopharynx (**Fig. 3**).

The patient underwent resection of the tumor by a lateral pharyngeal approach on January 29, 1998. The specimen excised at surgery showed a highly cellular tumor with dense spindle shaped cells arranged in various patterns including fascicular and storiform patterns. However, the storiform pattern was more frequent and mitosis was observed maximally in one cell per every 10 high power fields in this specimen (**Fig. 4**). The histopathological diagnosis was then changed to MFH. On February 18, 1998, the patient underwent an additional

surgery for the resection of the pyriform sinus by partial hypopharyngectomy while preserving the larynx with reconstruction using a free jejunal flap. However, the circulation of the free jejunal flap failed on the seventh post-operative-day due to a cervical infection. Reconstructive surgery using a pectoralis major flap was then performed. He started to swallow and phonate on March 10, 1998. For more than 4 years postoperatively, there has been no evidence of local recurrence or distant metastasis, as shown by a CT scan of the neck and a chest X-ray photogram.

### Discussion

MFH is a soft tissue sarcoma occurring mostly in middle-aged or older adults, especially arising from the proximal end of the extremities and retroperitoneum<sup>1</sup>. MFH is uncommon in the head and neck region, the frequency of MFH in the head and neck being only 3~10% of all MFHs<sup>1-3</sup>. Only 3 cases of MFH of the pharynx in adults have been reported so far<sup>4-6</sup>. One of the patients, with MFH of the oropharynx, underwent trans-oral excision, and the other two, with MFH of the hypopharynx, were treated by pharyngolaryngectomy or radiotherapy. There have been no reports of young adult patients with MFH, especially in the hypopharynx.

Pathologically, MFH is composed of an admixture of spindle-shaped fibroblastic tumor cells and bizarre mononuclear histiocytic tumor cells arranged in storiform pattern and some multinucleated giant cells. However, there are four<sup>7</sup> or five<sup>1</sup> different histological patterns of MFH: storiform-pleomorphic, myxoid, inflammatory, giant-cell<sup>7</sup>, and angiomatous<sup>1</sup>; therefore the diagnosis of MFH from biopsy specimens is thought to be difficult. Such specimens are not always large enough for an accurate pathological diagnosis, especially in tumors of the head and neck region. Moreover, inflammation of the pharyngeal specimens due to mechanical stimulation upon movement of the larynx can further confuse the diagnosis made from small biopsy specimens. It has been reported that biopsy specimens of MFH in the head and neck region were initially diagnosed as fibrosarcomas, or osteosarcomas<sup>8,9</sup>. In a case of

pharyngeal MFH, it was reported that the diagnosis at biopsy was rhabdomyosarcoma<sup>6</sup>. Our case was initially diagnosed as BFH from small biopsy specimens without high cellularity and mitosis. However, the excised specimen showed highly cellular proliferation of spindle cells and mitosis and clearly differed from that of the biopsy specimen. Moreover, the grade of malignancy in this particular MFH was low, this being also one of the reasons why the initial diagnosis was BFH.

Surgery is the treatment of choice for MFH since radiotherapy and chemotherapy are not effective<sup>10,11</sup>. Excision with wide margins is necessary since tumors are characterized by a high rate of local recurrence varying from 44~73%<sup>1,12</sup>. After the initial diagnosis of BFH, excision of the lesion without a wide margin was performed. Following the final pathological diagnosis a wide resection was performed but with preservation of the larynx.

In MFH, metastases most commonly occur in the lungs and lymph nodes<sup>1,2,13</sup>. Therefore CT scan of the chest and neck and ultrasonic examination of the neck are very useful for the follow up. It was reported that the tumor size, depth and inflammatory component influences the rate of metastasis<sup>1</sup>. In our particular case, as the tumor was small in size and was located in the pyriform sinus wall, there was no occurrence of metastasis. Blitzer et al.<sup>13</sup> stated that the prognosis of MFHs in the head and neck worsened with age, with mortality increasing from 13% at ages 0~19 years, to 72% at 60 years or older. Survival rates of more than 3 years were reported to be about 60%. Barnes et al. stated that the angiomatoid and myxoid types had a better prognosis while the giant-cell type was the worst<sup>2</sup>. Previous reports with pharyngeal MFH were followed up for a period of 2 years, but the long-term survival rates of these cases are unclear. Our young adult case of MFH with low grade malignancy in has had a long-term survival.

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