Radiation-induced Malignant Fibrous Histiocytoma of the Maxillary Sinus in a 25-year-old Patient with Nasopharyngeal Carcinoma: Early MR Imaging Characteristics

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The early MR imaging characteristics of a malignant fibrous histiocytoma (MFH) in a 25-year-old man under the radiation treatment of nasopharyngeal carcinoma were presented in this study. The tumor developed four years and 9 months following the 3-dimentional conformal radiotherapy of nasopharyngeal carcinoma. Early magnetic resonance imaging features were nonspecific for neoplasm and not distinguishable from mycetoma. Final diagnosis of MFH was achieved based on histopathology and immunohistochemical stains. The patient underwent lateral rhinotomy and medial maxillectomy with the en bloc resection of the tumor with an uneventful postoperative course.

Key words: Malignant fibrous histiocytoma; Magnetic resonance imaging; Maxillary sinus, Nasopharyngeal carcinoma, Radiotherapy

Malignant fibrous histiocytoma (MFH), a rare tumor originating from the histocytes [1], seldom arises from the maxillary sinus [2]. If occurs, it is usually mistaken as either sinusitis or dental problems in the beginning and not diagnosed until it results in bone destruction and invades the adjacent structures such as nasal cavity, orbit, or cheek. Here we present a case of maxillary MFH, which mimics a mycetoma on magnetic resonance (MR) images.

CASE REPORT

In August 1999, a 19-year-old man visited our hospital with the chief complaint of having suffered from epistaxis and a long-term nasal stuffiness with yellowish nasal discharge for 10 years. Physical examination revealed a protruding mass in the nasal choanal cavity with some blood clots on the surface of the mass. Contrast-enhanced MR study disclosed a large enhancing tumor occupying the entire nasal choanal cavity and the nasopharynx. Some enlarged nodes in the ipsilateral retropharyngeal space and the jugulo-digastric chain were detected. The right mastoid air cells were filled with fluid secondary to obstruction of the ipsilateral Eustachian tube. The bilateral maxillary sinuses showed submucosal swelling with smoothly enhancing mucosa. Biopsy of the nasal choanal mass was done, disclosing an undifferentiated carcinoma based on histopathology. Chest radiography, abdominal sonography, and whole-body bone scan did not disclose distant metastasis. The final tumor stage was T3N1M0, WHO type III.

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Afterwards, he received both radiotherapy and chemotherapy. 3-D conformal radiotherapy had been given for 2 months with a radiation dose of 7020 cGy delivering to the nasopharynx and 5400 cGy delivering to the lower neck. Chemotherapy included intravenous infusion of cisplatin 186 mg in each course for a total 7 courses and 5-FU (fluorouracil) 1680 mg each in the last four courses. Fibrotic scarring of the middle and inferior nasal turbinates and the floor of the nasal cavity occurred and progressed gradually since 2000. Functional endoscopic sinus surgery with bilateral inferior turbinectomy was performed for treatment of the choanal stenosis.

A series of MR and computed tomographic (CT) studies were undertaken to evaluate the treatment response of the nasopharyngeal carcinoma, showing vanishment of the nasopharyngeal tumor. In February 2002, the bilateral maxillary sinuses showed submucosal swelling with smooth mucosal enhancement. In November 2003, the enhancing mucosa of the right maxillary sinus became irregular and spiculated (Fig. 1a, 1b). We observed a small nodular lesion on the same location in July 2004. The nodule appeared isointense on T1-weighted images and hypointense on T2-weighted images and enhanced strongly after intravenous injection of gadolinium-DTPA (Fig. 1c, 1d). Such imaging presentation is favorable to fungal sinusitis with mycetoma. The patient experienced bloody nasal discharge half year later. In January 2005, biopsy of the nasal cavity mass revealed high grade pleomorphic sarcoma. CT study performed two weeks after the

![Figure 1. Axial MR images at the level of nasopharynx and maxillary sinuses. A small patch of irregular tissue develops in the posteromedial mucosal layer of the right maxillary sinus, showing heterogeneous signal intensity on T2-weighted image (a) and irregular enhancement on post-contrast T1-weighted image (b) four years after radiotherapy. It forms a small nodule which is hypointense on T2-weighted image (c) and strongly enhances on T2-weighted image (d) 8 more months later.](image-url)
biopsy disclosed a nasal cavity mass, which was the extension of the original maxillary sinus mass via the destroyed medial sinus wall (Fig. 2).

The patient was treated by lateral rhinotomy and medial maxillectomy with total resection of the tumor. Immunohistochemical stains showed that the tumor cells were positive for vimentin, but negative for muscle actin, S-100, CD34 and cytokeratin, leading to the diagnosis of pleomorphic malignant fibrous histiocytoma (Fig. 3).

**DISCUSSION**

MFH occurs most commonly in the extremities, followed by abdominal cavity and/or retroperitoneum. It seldom develops on the head and neck region [3] and is extremely rare in the maxillary sinus [2]. As other maxillary neoplasm, maxillary MFH is usually mistaken as maxillary sinusitis, osteomyelitis or dental problems due to the non-specific clinical presentations, including nasal obstruction, facial soft tissue swelling, pain, and/or visual disturbance. If unrecognized, it is often treated by antibiotics, sinus drainage or dental extraction in the early stage. The mysterious symptoms and signs postpones the diagnosing procedure and accurate diagnosis until the development of bone destruction, teeth loss, invasion of the nasal cavity, cheek, or orbital structures [4-6].

In the past, the role of radiography in the early detection of maxillary neoplasm is limited. Plain radiography, including the panoramic, intraoral, and Water’s radiographs, is unable to distinguish neoplasm from sinusitis based on the soft-tissue opacification of the maxillary sinus. The plain radiography detects the malignant sign of the neoplasm according to the destruction of the sinus wall, orbital rim, or alveolar bone. CT study provides excellent spatial resolution superior to plain radiography, and therefore, has a better opportunity to detect the maxillary neoplasm earlier. Unfortunately, a review of the literature shows that CT study does not bring earlier detection of the maxillary MFH [5, 7-10]. It is believed that the indolent clinical symptom might delay the CT imaging study. The MR imaging features of the MFH were first described in 1989 by Mahajan H. et al [2]. In 39 reported cases, the tumor arose from the maxillary sinus in one case (2.56%). Generally, MFH shows intermediate signal intensity on T1-weighted images, heterogeneous hyperintense on T2-weighted images [2, 11] and shows heterogeneous enhancement on contrast-enhanced T1-weighted images [12]. However, the MR imaging features were neither specific nor pathognomonic for MFH.

Nasopharyngeal carcinoma (NPC), a rare malignancy in the west but quite common in the east especially in southern China and Taiwan, is a curable disease under the treatment of radiotherapy with...
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a overall 5-year survival rate as high as 65.5% to 70.6% [10, 13]. The radiation dose used in the treatment of NPC is high, reaching 70 Gy to 80 Gy, commonly leading to osteoradionecrosis of the skull base and temporal bones, and radiation necrosis of the temporal lobes of the brain. As long as the prolongation of the life span after radiotherapy, the NPC survivors might encounter radiation-induced sarcomas in the head and neck region. A prevalence of 0.38% of the long-term survivors of radiation-induced MFH was documented in a study of 3223 NPC patients in Taiwan [10]. The estimated dosage of radiation exposure of the nasopharynx ranged from 50.4 Gy to 80.1 Gy. In their study, MFH occurred in the maxillary sinus in seven cases. The tumors had already caused bone destruction, tumor invasion of the infratemporal fossa, nasal cavity, oral cavity, inferior orbital fissure, and cavernous sinus at the time of CT imaging. Local recurrence occurred in all cases with 9 months and 75% of cases expired within 30 months [10].

In our case, the dosage of radiation delivering to the maxillary sinuses was 6640 cGy. The duration from radiotherapy to the detection of the maxillary nodule was four years and 9 months. The nodular lesion resembles a mycetoma on MRI, showing hypointense on T2-weighted images and strong enhancement [14]. Furthermore, the MR imaging findings in our patient were different from the typical features of MFH reported by Mahajan H. et al, and Munk et al [2, 11]. To the most of our knowledge, the atypical early MR imaging characteristics of maxillary MFH of a NPC survivor have not been documented before.

From this case study, we learn that regular MR studies provide a possibility for early detection of radiation-induced maxillary MFH. However, the MR imaging features of maxillary MFH are not pathognomonic and should be distinguished from mycetoma. The early diagnosis relies on high index of suspicion and knowledge of the clinical history of high-dosage radiation exposure, timing from the radiation to occurrence of tumor development, regular MR imaging follow-up studies, and early histological analysis. The role of MR imaging studies is not only to monitor the response of the nasopharyngeal carcinoma to the radiotherapy and disease progression, but also to delineate the early change of the sinus mucosa being exposed to radiation, occurrence and progression of the tumor, spatial relationship of the tumor and adjacent structures, and prompt early biopsy and histological analysis.

REFERENCE

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我們在此報告一位二十五歲因鼻咽癌接受放射線治療後併發上頜竇之惡性纖維組織細胞瘤
之早期磁振造影影像的特徵。該腫瘤發生於放射線治療四年又九個月之後。早期磁振造影影像
的特徵並無法與足菌腫作鑑別診斷。最後的確定診斷有賴於組織病理切片以及免疫螢光染色等
檢查。此病患接受外側鼻腔及內側上頜竇切除後至今未曾覆發。

關鍵詞：惡性纖維組織細胞瘤，磁振造影影像，上頜竇，鼻咽癌，放射線治療