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Undifferentiated sarcoma of the maxillary sinus: Report of a rare case in an adult

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Abstract

A 51-year-old man complained of left facial swelling and recurrent nasal bleeding. A giant solid tumor in the left maxillary sinus was detected on head CT and MRI, and this tumor was destroying the maxilla and extending into the orbit, pterygoid muscle and posterior paranasal sinuses. The resected specimen consisted of spindle cells containing necrotic material. Histological examination revealed immature tumor cells, and immunohistological study of the tumor showed staining was only positive for vimentin. We accordingly diagnosed undifferentiated sarcoma in the maxillary sinus. Combination chemotherapy with vincristine, doxorubicin, cyclophosphamide/ifosfamide with mesna and etoposide was administered; however, the tumor was unresponsive and the patient died after around 3 months.

Keywords: Undifferentiated sarcoma; Maxillary sinus; Combination chemotherapy

1. Introduction

Undifferentiated sarcomas arising from the maxillary sinus has not been reported previously and respond poorly to chemotherapy and radiotherapy, with resultant poor prognosis. Sarcomas of the head and neck are usually rhabdomyosarcomas in children, while leiomyosarcoma and fibrosarcoma are more often seen in adults [1]. We treated an adult in whom we diagnosed undifferentiated sarcoma not otherwise classifiable arising in the maxillary sinus, and we report this case herein.

2. Case report

On 22 November 2004, a 51-year-old man with the left cheek swelling and recurrent epistaxis for a week was referred to our hospital. Computed tomography of the head demonstrated expansile, osteolytic destruction of the left maxilla and associated soft tissue densities in the left orbit.

Physical examination showed the left hard and painful facial swelling and epiphora. However, eye movement was normal (Fig. 1). The hard palate around the gingiva was irregular and reddish in areas. A friable tumor was seen expanding from the left nasal cavity to the nasopharynx, and the tumor was biopsied. Computed tomography (CT) and magnetic resonance imaging (MRI) of the head demonstrated a large mass measuring about 8 cm in diameter in the left maxillary sinus invading the skullbase (cavernous sinus), the left orbit, nasopharynx, buccal soft tissues, infratemporal fossa, and oral cavity with massive osteolytic destruction (Fig. 2a and b). CT of the neck and the thorax were unremarkable.

Macroscopically, the biopsy specimen showed areas of marked necrosis and discharge. Routine light microscopy revealed small regular spindle-shaped cells with scanty cytoplasm and hyperchromatic spindle-shaped nuclei. These cells demonstrated diffuse proliferation. However, tumor

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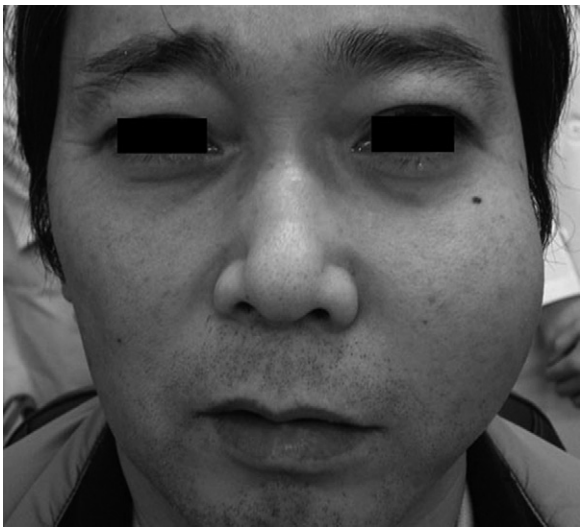

Fig. 1. The left cheek swelling, centered around the maxillary sinus was shown.

cells exhibited very little interstitial fibrillar collagen and minimal storiform or whorl patterns, although myxoid stromal elements were seen. Abnormal mitotic figures were present in each microscopic field. These characteristics were indicative of sarcoma (Fig. 3). Immunohistochemical analysis showed positive staining for vimentin(mesenchymal tissue marker), and negative staining for epithelium antigenBer-Ep4, AE1/AE3, CK7, CK20, and 34βE12 (epithelium), desmin(muscle), SMMC-1(smooth muscle), and HHF35(striated muscle) (Fig. 4a and b), CD34(hemangioendothelial cell), S-100 protein(nerve fiber), and HMB45(melanoma cell). The pathological diagnosis was undifferentiated sarcoma. Therefore, we diagnosed the tumor as an unclassifiable, undifferentiated sarcoma.

The patient was taken the chemotherapy according to the protocol used to treat rhabdomyosarcoma, extraosseous Ewing's sarcoma, and undifferentiated sarcoma [2,3]. This chemotherapy regimen comprised vincristine (1.5 mg/m^2), doxorubicin (37.5 mg/(m^2 day)), cyclophosphamide (600 mg/(m^2 day)) for 2 days with mesna (360 mg/m^2) for five doses, ifosfamide (1800 mg/(m^2 day)) for 5 days with mesna (360 mg/m^2) for five doses, and etoposide (100 mg/(m^2 day)) for 5 days. After about a month (when the first course of chemotherapy had been completed), severe side effects such as nausea, vomiting, and high fever (39–40 °C) developed. Chemotherapy was therefore not resumed. Enlargement of the maxillary tumor and new lymphadenopathy in the neck were evident, with recurrent bleeding of the palate. Emergent tracheotomy was performed due to severe dyspnea. The patient died on 18 March. However, autopsy was not performed.

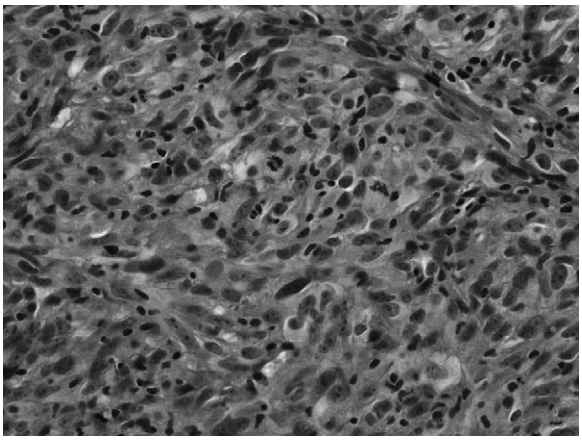

Fig. 3. Light microscopic examination revealed that it was composed of small regular spindle-shaped cells with scanty cytoplasm and hyperchromatic spindle-shaped nuclei. The cells demonstrated diffuse proliferation (HE, 400×).

3. Discussion

Most malignant tumors of maxillary sinus are squamous cell carcinomas or sarcomas. In children such sarcomas are

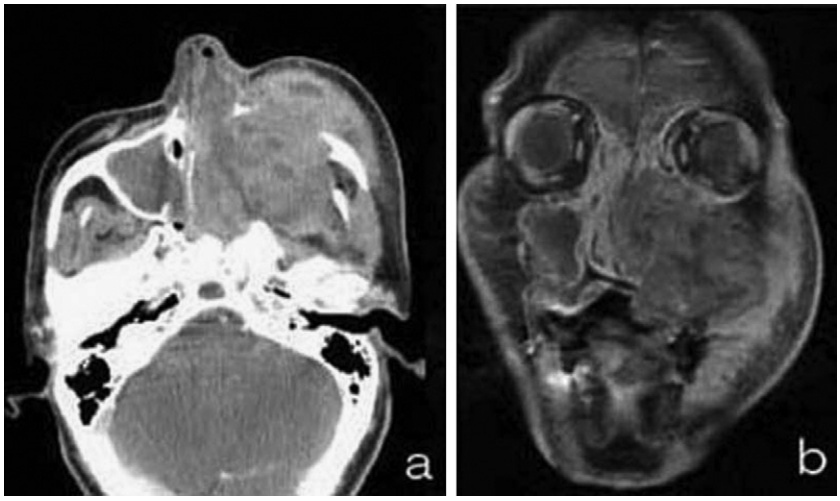

Fig. 2. (a) CT and (b) MRI showed that the tumor had destroyed the some of the maxilla, right ethmoid sinus, sphenoid sinus, and that it extended into the right nasal cavity, the orbit, pterygoid muscle, and the skullbase involved.

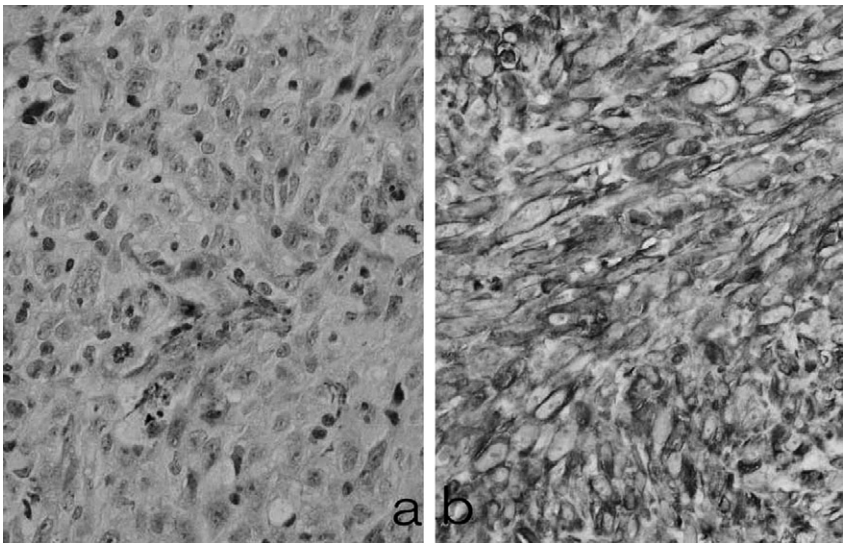

Fig. 4. (a) Immunohistochemical staining demonstrated positive for vimentin, confirming the mesenchymal origin of this tumor (400×). (b) Immunostaining for HHF35, which is characteristic in smooth muscle and cross-striations of striated muscle, was absent (400×).

usually rhabdomyosarcomas, while in adults, leiomyosarcoma or fibrosarcoma are more frequent.

Malignant fibrous histiocytoma (MFH) or spindle cell carcinoma (SPCC) are tumors about which much remain unknown. The cell origin, molecular mechanism of pleomorphism and mechanism of pleomorphic change in a cell undergoing malignant change have not been elucidated. MFH-like histological changes are observed in many soft tissue tumors, and malignant lymphomas. These changes occur in many subtypes of sarcomas such as osteogenic sarcoma, chondrosarcoma, leiomyosarcoma, rhabdomyosarcoma, and liposarcoma. MFH has been regarded as one of tumor classification from its special histopathological features. In clinical pathological studies these tumors are divided into low-grade fibrous tumors and fibrous histiocytic tumors. With the establishment of pathological diagnostic methods, MFH-like histological features can be seen in changes in cellular differentiation of many sarcomas. Although the careful review of H&E stained sections remains of critical and central importance in this evaluation, the recent improvement in the immunohistochemical diagnostic techniques may add diagnostically valuable information.

Undifferentiated sarcoma of the liver has been reported [4]. The microscopic characteristics composed of small spindle-shaped cells, with scanty cytoplasm and hyperchromatic irregular shaped nuclei, demonstrating diffuse proliferation. Immunohistochemical evaluation did not any positive reaction without PAS. The pathological findings of this patient were similar to those of undifferentiated sarcoma of the liver.

Extra immunohistochemical analysis showed negative staining for CK7, CK20, CK34βE12, CK(CAM5.2), epithelium antigen(Ber-Ep4), EMA, claudin-4, and α-SMA(1A4). This patient was diagnosed as undifferentiated sarcoma. It seems to be originated from masenchymal components aberrant tissue or embryonal remnants.

Pathologic findings in this case showed undifferentiated sarcoma, not further classifiable. Immunohistochemical with some specific antibodies, have proven to be the only definitive method to identify this variant sarcoma. We are not aware of cases with similar pathological findings reported in English literatures. To our knowledge, this report is the first of such sarcoma arising from the maxillary sinus. This patient was diagnosed as undifferentiated sarcoma. It seems to be originated from mesenchymal components of aberrant tissue or embryonal remnants.

We based on the protocol for Stage 1 (T1bN1M0), Group IV of the Intergroup Rhabdomyosarcoma Study (IRS) III [5] because the specific chemotherapy protocol for this sarcoma was not established. However, during a month of chemotherapy, the tumor enlarged rapidly and the new neck lymphadenopathy developed.

In the IRS trials, rhabdomyosarcomas have been subdivided into orbital and nonorbital types based upon the primary tumor lesion. The nonorbital types are further subdivided into parameningeal (arising from the nasopharynx, paranasal sinus, external ear canal, middle ear, mastoid, and infratemporal fossa) and nonparameningeal types [6]. In the IRS series, 11% of all head and neck rhabdomyosarcomas were found to be nonorbital and nonparameningeal. The anatomic disposition of RMS is a significant prognostic indicator, because parameningeal type has a worse prognosis than nonparameningeal type [7,8].

The grade of malignancy according to the French Federation of Cancer Centres (histopathological grading of soft tissue sarcomas) [9] showed that the present tumor scored 3 for differentiation, 3 for mitosis count, and 1 for tumor necrosis, giving a total score of 7. This score suggests high grade malignancy.

The present protocol was developed to increase the response and survival rates of patients with rhabdomyosarcomas,

extraosseous Ewing's sarcoma, and undifferentiated sarcoma, all of which have poor prognoses.

The development of effective and increasingly intensive and effective multimodality combination regimens in the treatment of sarcomas has resulted in a marked increase of survival time and cure rate. However, for the majority of patients with stage IV rhabdomyosarcomas, the cure rate has remained at a disappointing level of 20–30% [5]. In addition to stage at diagnosis, primary lesion and histology of the tumor are important prognostic factors.

The present tumor was unresectable because of the invasion into the cavernous sinus and resistant to treatment with the above-mentioned combination chemotherapy, resulting in the poor prognosis.

We reported the first case of undifferentiated sarcoma, not further classifiable, arising from the maxillary sinus. This sarcoma responds poorly to chemotherapy with resultant poor prognosis.

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