Case Report
Myeloid sarcoma of the cheek and the maxillary sinus regions

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Abstract
Myeloid sarcoma (MS) is a rare, extramedullary malignant tumor composed of immature myeloid precursor cells and myeloblast. Most MSs occur in the subperiosteal region of the bone, with the skull, sternum, ribs, and proximal portions of the long bones being the common sites of involvement. It is thought that the MS tumor originates in the bone marrow, and traverses the Haversian canals to reach the subperiosteum. Various reports have also described the involvement of the liver, spleen, brain, heart, pharynx, uterus, vagina, skin, kidney, and other soft tissues in the formation of the tumor.

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1. Introduction
Although myeloid sarcomas (MSs) are not commonly found in the oral cavity, the formation of such tumors can involve the palate, gingiva, and cheek. Herein, we present a report of MS without hematological disorders, and discuss our experience with this specific clinical finding as well as our surgical approach.

2. Case report
A 56-year-old man suffered from a swelling (lesion) in the left cheek with a noticeable mass for months. This cheek lesion slowly enlarged over the months and a mild tenderness was reported. He visited an oral surgery department, where a physical examination was performed, which revealed a soft and solid mass, measuring 4 cm in diameter on the left cheek.

The hematocrit level of the patient was 48%, and his white blood cell count was 9500/mm³ with 13.5% lymphocytes and 81.2% polymorphonuclear neutrophilic granulocytes. His treatment involved incision and drainage as well as administration of oral antibiotics; however, the swelling did not subside following the treatment. Therefore, a pathological analysis was conducted, which revealed a granulation tissue without malignancy, prompting a referral to the laryngological service for further evaluation. A computed tomography scan was performed, which revealed an enhancing soft tissue mass (diameter: 4.2 × 4.0 × 3.7 cm) involving the left cheek and the left maxillary sinus region. The differential diagnosis could be an immature abscess, a lymphoma, and several other types of tumors (Fig. 1). After having a discussion with the patient, we performed an operation to remove the lesion on the left cheek and the left maxillary sinus region, by following the Caldwell–Luc surgical procedure. A tissue proofing was carried out, which revealed that the specimen was composed of brown tissues diffusely infiltrating the left cheek without epithelial rupture and measuring up to 3 × 2.5 × 1.2 cm. Further examination of the tissue sections showed diffuse infiltration of tumor cells in the fibroadipose tissue and skeletal muscle fibers. The tumor cells were medium sized, with oval and vesicular nuclei, open chromatin pattern, and some with a prominent nucleolus. At higher magnification levels, the tumor cells exhibited a high nuclear-to-cyttoplasmic ratio, with round- to ovoid-shaped nuclei.

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An immunohistochemical analysis revealed that the neoplastic cells were positive for CD34, CD45, CD56, CD117, and myeloperoxidase (MPO), but not for CD3, CD4, CD20, CD30, CD68, terminal deoxynucleotidyl transferase (TdT), vimentin, or cytokeratin AE1/AE3 (Fig. 2). The results of these diagnostic tests and examination suggested MS. The patient was subsequently referred for a hematological workup, in order to exclude other synchronous myeloproliferative disorders. No alterations in the peripheral blood profile were noted, and the results of his bone marrow biopsy showed normocellular marrow without malignancy. Immunohistochemical staining with anti-CD34 showed scant positive cells. No CD117-positive cells were identified, excluding acute myeloid leukemia. On the basis of the composite clinical-pathological data, the patient accepted combination chemotherapy of low-dose cytarabine (arabinofuranosyl cytidine) (20 mg/day subcutaneously, on days 1–25) and aclarubicin (10 mg/day intravenously, on days 1–4). Four months after admission, the cheek lesion had completely regressed and the patient was in clinical and hematologic remission (Fig. 1). We did not supplement the course of treatment with radiation therapy because the disease did not persist after the combination chemotherapy was completed.

3. Discussion

MS is a localized, solid, extramedullary tumor composed of immature myeloid precursor cells. Burns first described a patient with proptosis and green retro-orbital tumors in 1823, and Dock later established the relationship of MS with acute leukemia. Turk et al reported the first case of myelocytic leukemia associated with chloroma. Almost all cases of chloroma reported since then have been shown to be associated with myelocytic or monocytic leukemia. The MS tumor is more common in children and young adults. A vast majority of MSs occur in the subperiosteal region of the bone, with the skull, sternum, ribs, and the proximal portions of long bones being common sites of involvement. MS is defined as a tumor mass of myeloid blasts with or without maturation occurring at an anatomical site other than the bone marrow, with normal architectural effacement. It is particularly important to identify any destructive growth to characterize the lesion to be MS in patients with leukemia.

The existing literature suggests that the tumor arises from the bone marrow, and traverses the Haversian canals to reach the subperiosteum. In our case, the tumor (lesion 4.2 × 4.0 × 3.7 cm) occurred on the left cheek and the left

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Fig. 1. (A) Enhancing soft tissue (diameter of 4.2 × 4.0 × 3.7 cm) involving the left cheek and the left maxillary sinus region. (B) The cheek lesion completely regressed after therapy.
MS is usually composed of relatively uniform, primitive cells that may be peroxidase positive or negative. Ruppaport et al\(^9\) maintains that a definite diagnosis must be based on the identification of such cells in the blood and marrow. The diagnosis becomes difficult when nonpigmented MS precedes acute leukemia. One form of the disease, reticulum cell sarcoma, occurs primarily as a single or multiple solid tumors. Another more malignant form of the disease, acute myelocytic or monomycytic leukemia, commonly occurs as a widespread infiltrative malignancy without discrete tumor formation. An intermediate between these two extremes may be an MS, a phase of widespread solid tumors. The use of only four antibodies (MPO, CD68, lysozyme, and CD34) has been proposed to distinguish the more common variants of MSs. A study of 30 cases showed CD117 reactivity in 87%, MPO, 97%; lysozyme, 93%; CD34, 47%; CD45, 84%; CD43, 97%; TdT, 37%; CD79a, 20%; CD20, 10%; CD3, 10%; and CD10, 1%. There is no standard treatment for MS, especially when it occurs de novo\(^9\); rather, treatment is similar to that of a systemic disease, as it is considered to be a manifestation of such diseases. When a patient with MS has leukemia or relapsed leukemia, combination chemotherapy for acute leukemia may induce complete remission. Additional radiation therapy is often considered when the disease persists after chemotherapy. Systemic chemotherapy also may induce complete remission in the patient with de novo MS.\(^10\)

The prognosis of MS is generally unfavorable. Pileri et al\(^8\) reported that of the 67 patients for whom follow-up data were available, 60 died from the disease (89.5%). Interestingly, all the seven survivors achieved complete remission of their MS following first-line therapy, in contrast to only eight of the 60 patients who died achieved complete remission. Bone marrow transplantation resulted in longer survival than conventional therapy. Based on these results, the authors strongly recommended that patients with MS should undergo high-dose therapies as a front-line approach, noting that this appeared to be the only chance to achieve complete remission and to cure the disease. Even though the prognosis of MS is very poor, it should be diagnosed as early as possible because early high-dose therapies offer the only possibility of cure. Finally, in patients with leukemia, even in those in complete remission, physicians should not overlook symptoms mimicking facial cellulitis or abscess and must bear in mind the possibility of MS.

We present this case of specific radiological finding and surgical approach to MS of the cheek and maxillary sinus. The disease is often easily misdiagnosed, so we recommended all cases of head and neck tumor refractory to medical treatment be treated with aggressive surgical approach for complete tissue proof.

In conclusion, it is remarkable that in clinical practice there is frequent misdiagnosis of MS. Histological analyses show that MS has an abundance of monomorphic polyhedral cells with irregular nuclear contours. Immunohistochemical studies may help in reaching a definitive diagnosis because myeloid cells are reactive to antibodies against lysozyme, myeloperoxidase, and chloroacetate esterase. In addition, flow cytometry and cytogenetics may help to improve the likelihood of a correct, definitive diagnosis. Imaging studies can also be useful to rule
out inflammatory lesions. Further clinical hematological investigations, including a bone marrow biopsy, are therefore recommended in each case of MS. MS of the head and neck carries prognostic significance with blast crisis, so an aggressive surgical approach for complete tissue proof is suggested for recurrent lesions that are refractory to medical treatment. Complete excision of tumor with a satisfactory free margin can more effectively create the foundation for a reliable and accurate diagnosis for suspected malignancy, and better differentiate it from infection etiology.

References