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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.

Keywords: cheek; maxillary sinus; myeloid sarcoma

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/mm^3 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-cytoplasmic ratio, with round- to ovoid-shaped nuclei.

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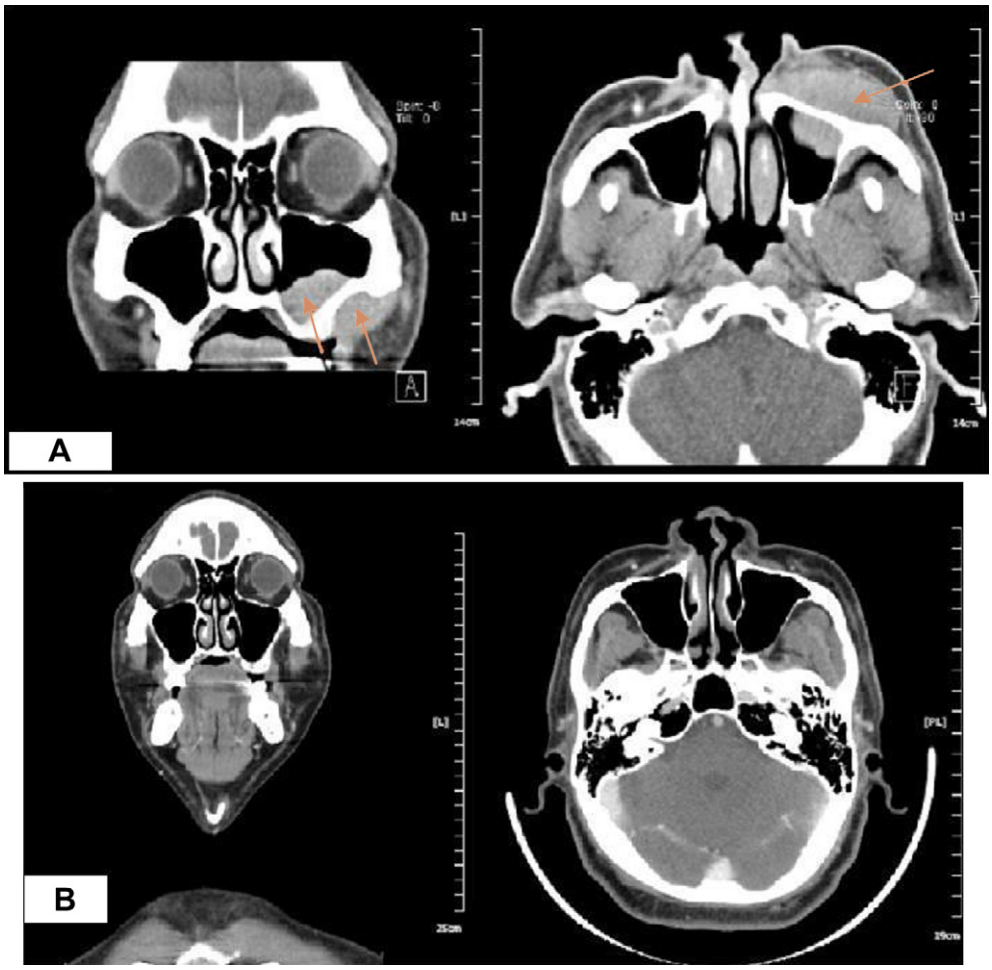

Fig. 1. (A) Enhancing soft tissue (diameter of 4.2 × 4.0 × 3.7 cm) involving the left cheek and the left maxillary sinus region. (B) The cheek lesion completely regressed after therapy.

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 clinicopathological 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[1] first described a patient with proptosis and green retro-orbital tumors in 1823, and Dock[2] later established the relationship of MS with acute leukemia. Turk et al[3] 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.[4] 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.[5] 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.[5] In our case, the tumor (lesion 4.2 × 4.0 × 3.7 cm) occurred on the left cheek and the left