Rare Presentation of Myeloid Sarcoma: Nasopharyngeal Myeloid Sarcoma

Yetkin Zeki YILMAZ[1], Atakan DEMİR[1], Fırat TEVETOĞLU[1,*],
Ayşegül BATIOĞLU KARAALTIN[1]

Abstract

Myeloid sarcoma is a rare tumor of extramedullary immature myeloid cells and very few cases of nasopharyngeal involvement have been documented. Despite being most common in skin, bone and lymph nodes; it can be seen in any region with nasopharynx being one of the rare sites. In this article, we aimed to present a case of nasopharyngeal myeloid sarcoma. 58 – year – old male who was previously diagnosed with AML transformed from MDS and treated with chemotherapy, presented with complaints of hearing loss in the right ear and swelling in the right side of the neck. He had a mass lesion in the right half of the nasopharynx in the endoscopic examination performed upon the presence of hearing loss and serous otitis findings. Imaging findings revealed a mass lesion starting from the right half of the nasopharynx extending to the posterior of the carotid space and paravertebral area. Patient was diagnosed with acute myeloid leukemia (AML) transformed from myelodysplastic syndrome (MDS) and was treated with chemotherapy regime consisting of cisplatin and concomitant radiotherapy. Since myeloid sarcoma is a rare tumor seen in 3-8% of patients with AML, high suspicion and immunohistochemical analysis are important in the diagnosis of this tumor.

Keywords: *nasopharynx; myeloid sarcoma; acute myeloid leukemia.*

[1]Istanbul University – Cerrahpaşa , Cerrahpasa Medical Faculty, Department of Otorhinolaryngology Head and Neck Surgery, İstanbul- Turkey
* Corresponding author: firattevetoglu@gmail.com; ORCID: 0000-0002-1744-6995

Yetkin Zeki YILMAZ: yetkin.yilmaz@iuc.edu.tr; ORCID: 0000-0002-5734-9751
Atakan DEMİR: datakan1995@gmail.com; ORCID: 0000-0002-7867-5185
Ayşegül BATIOĞLU KARAALTIN: batioglu@yahoo.com; ORCID: 0000-0002-8620-4375
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Introduction

Myeloid sarcoma is a pathological definition expressing the extramedullary proliferation of blasts in one or more myeloid series. It usually occurs as an isolated extramedullary leukemic tumor or as a relapse of AML(1). Although this tumor most commonly occurs in the skin, bone and lymph nodes; it can be seen in many regions such as the CNS, oral and nasal mucosa, breast, genitourinary tract, chest wall, pleura and mediastinum(2) . The most common clinical findings are severe pain due to the mass effect of the tumor and abnormal bleeding. Treatment options such as chemotherapy, radiotherapy, hematopoietic stem cell transplantation and targeted therapy are available. The prognosis is worse for those that occur in presence of AML(3).

In this article, we aimed to present a case of nasopharyngeal myeloid sarcoma, which is a rare presentation of myeloid sarcoma.

Case Presentation

A 58 –year – old male patient, who was previously diagnosed with AML transformed from MDS and treated with chemotherapy, presented with complaints of hearing loss in the right ear and swelling in the right side of the neck. Endoscopic examination revealed serous otitis findings in the right ear and a submucosal mass lesion in the right half of the nasopharynx. Cranial and cervical computed tomography (CT) and magnetic resonance imaging (MRI) of the patient revealed a mass lesion starting from the right half of the nasopharynx extending to the posterior of the carotid space and paravertebral area, surrounding the internal carotid artery by 360 degrees, penetrating into the carotid canal (Figure 1). In the PET-CT scan of the patient, a hypermetabolic mass lesion involving bilateral nasopharynx, being more prominent on the right was thought to be due to the primary disease (Figure 2). In complete blood count, white blood cell count was 7900/microliter with %52.1 neutrophils, %37.5 lymphocytes, hemoglobin was 15.1 g/dl and platelet count was 222000/microliters. While nasopharyngeal carcinoma was the most possible diagnosis, histopathology and immunohistochemistry of the punch biopsies taken from the nasopharyngeal mass revealed myeloid sarcoma. For the treatment, chemotherapy regime consisting of cisplatin and concomitant radiotherapy were initiated by the

medical oncology and radiation oncology departments but the patient died 1 week after the beginning of the treatment regime.

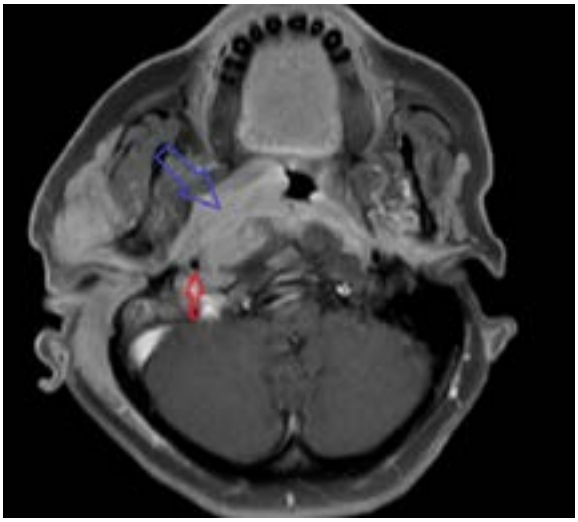

Figure 1. *Contrast-enhanced neck magnetic resonance imaging axial section revealed a mass lesion starting from the right half of the naso-pharynx (indicated by blue arrow) and surrounding the internal carotid artery (indicated by red arrow) by 360 degrees.*

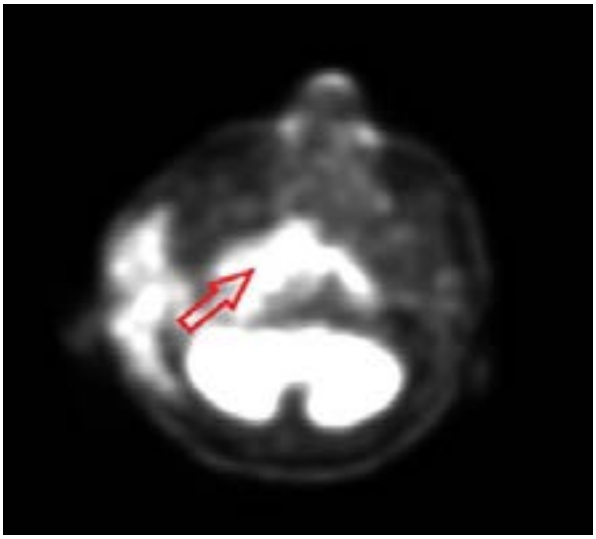

Figure 2. *PET-CT scan axial section indicated a mass lesion at the skull base, involving bilateral nasopharynx (indicated by red arrow).*

Discussion

Myeloid sarcoma is the extramedullary proliferation of blasts in myeloid series, usually occurring as an isolated extramedullary leukemic tumor or in association with other myeloid disorders (mostly AML, myeloproliferative diseases especially CML and MDS) (1). It is a rare tumor seen in 3-8% of patients with AML. Despite being most common in skin, bone and lymph nodes; it can be seen in any region with nasopharynx being one of the rare sites (2). Myeloid sarcoma is usually misdiagnosed initially due to its clinical presentation with nonspecific symptoms (4). Patients can present with sore throat, jaw pain, sinus pain, skin lesions (papules, nodules or rash), tonsillar enlargement or lymphadenopathy. In the oral cavity, authors have emphasized that myeloid sarcoma could mimic pyogenic granuloma, abscess or other inflammatory processes, thereby delaying biopsy and diagnosis (5).

The characteristic microscopic appearance of myeloid sarcoma is Indian mesh pattern. It is classified as granulocytic, monoblastic and myelomonocytic according to the most common cell type. The most common positive immunohistochemical markers are CD68 and CD13 (CD68 for macrophages and CD13 for granulocytes); myeloperoxidase (MPO) and CD117 (markers for myeloid differentiation) ; lysozyme (a marker of monocytic lineage), CD43 (on the myeloid cells as well as the T cells and the B precursors) , CD34 and TdT (expressed on immature cells) (1,3). Initially, nasopharyngeal carcinoma was suspected in our case due to the clinical findings and localization. The main symptoms of our case were unilateral hearing loss which was explained by serous otitis media and unilateral neck mass, which were highly consistent with nasopharyngeal carcinoma. However, submucosal appearance of the lesion was suggestive for diagnoses other than nasopharyngeal carcinoma. Hence, our case was diagnosed with myeloid sarcoma, in the context of AML history, the presence of lymphoepithelial component and the presence of CD117 positive cells.

In the case of nasopharyngeal myeloid sarcoma reported by Raphael et al., although high grade lymphoma is suspected primarily for a 73-year-old man presented with a right-sided nasopharyngeal mass confirmed by imaging; absence of expression of lymphoid markers and myeloperoxidase

lead the final diagnosis of MS. The patient was treated with conventional induction AML therapy. The clinical outcome was favorable, the PET-CT scan showed no abnormal FDG uptake, the bone marrow and blood count were normal except for a megakaryocytic hypoplasia. However, three months later, another bone marrow examination revealed an overt acute myeloblastic leukemia and the patient was enrolled in a phase I trial with the Etoposide. After 2 cycles there was no response, the treatment was stopped and patient died from disease progression (6). In another case report, further work-up was suggested to a 73 years old male patient who presented with one month history of dysphagia and noisy breathing. Hypertrophy of the lingual and palatine tonsils was detected in the endoscopic examination. Biopsies taken from both tonsils were compatible with myeloid sarcoma and the patient opted for treatment with palliative chemotherapy but he died due to the rapid progression of the disease (7)where a mass (tumour. In an article three cases of myeloid sarcoma were presented: (1) A 17-year-old boy with AML-M4 presented with sudden bilateral facial paralysis, sudden hearing loss, vision disturbance and a rapidly growing mass in bilateral postauricular area (2). A 17-year-old girl presented with ill-defined soft mass measuring 5 cm in diameter in the right preauricular area (3). 33-year-old man presented with multiple masses on his skin, bilateral cervical and axillary lymphadenopathy. All patients undergone chemotherapy and immunotherapy with interleukin-2 was also added for case 3. However, all of them relapsed and died (8). For our case, a second regime of chemotherapy consisting of cisplatin and concomitant radiotherapy was initiated as the treatment; yet patient died one week after.

Although there are different approaches in the treatment of the disease, such as chemotherapy, radiotherapy, hematopoietic stem cell transplantation and targeted therapies (imatinib, gemtuzumab, etc.), no clear consensus or recommendation exists in the literature. The prognosis seems more dismal than overt AML although it remains controversial. In the absence of leukemia, surgical removal of the tumor followed by local radiotherapy may be performed; because granulocytic sarcomas are generally regarded as radiosensitive tumors (9). Local radiotherapy may also yield better local tumor control if residual disease persists or the tumor relapsed after initial chemotherapy. Also, high-dose chemotherapy followed by stem cell

transplantation may be associated with a higher probability of survival or cure(10). 9 of 497 AML patients with isolated myeloid sarcoma retrospectively reviewed by Lee J.Y. et al. and the most common site for MS was head and neck region, with 4 patients evolving into AML in a median time of 13.4 months. Patients achieving complete remission after first-line treatment was higher in the "local treatment with or without systemic treatment (LS)" group than in the "systemic treatment only (S)" group and while all patients in the LS group survived, all those in the S group died (P=0.012) (11). Patients with myeloid sarcoma combined with AML have a poor prognosis. Even after chemotherapy with or without radiotherapy, as many as 85% of the patients relapse within 1 year (12) and these relapses are usually the cause of death (6–8).

Myeloid sarcomas are related with worse prognosis especially in the presence of AML. So, early treatment initiation is crucial, which makes differential diagnosis more important. For a nasopharyngeal mass, it should be highlighted that submucosal localization was not consistent with nasopharyngeal carcinoma and suspicious for other pathologies. In addition, history of hematological diseases such as myelodysplastic syndrome or myeloblastic leukemia is also suggestive for myeloid sarcoma. If myeloid sarcoma is one of the possible diagnoses, it is important to communicate with the pathologist to lead the diagnosis to a more accurate direction, in order to prevent waste of time.

Conclusion

In conclusion, myeloid sarcoma is a rare tumor seen in 3-8% of patients with AML and extremely rare when speaking of nasopharyngeal myeloid sarcoma in particular. High suspicion and immunohistochemistry are important for the diagnosis of this tumor having a poor prognosis. Local treatment options such as surgical removal and radiotherapy could be considered in isolated tumors in the absence of leukemia, but systemic chemotherapy would be the treatment of choice in patients with leukemia.

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