

# Granulocytic Sarcoma of the Larynx: Case Report

## Lareneal Granülositik Sarkom: Olgu Sunumu

**Mehmet YILMAZ, MD, Marlen MAMANOV, MD, Ahmet GÖKÇEL, MD**

İstanbul University Cerrahpaşa Medical Faculty, Department of Otorhinolaryngology Head and Neck Surgery, İstanbul

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### ABSTRACT

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Granulocytic sarcomas are localized extramedullary deposits of myeloid leukemia cells. These tumors are considered to be specific lesions of acute myeloid leukemia or the onset of a blast crisis in chronic myelogenous leukemia. Granulocytic sarcoma can occur anywhere in the body. Involvement of the larynx by hemopoietic tumors is considered as a rare event. The predominant symptoms are difficulty of breath and hoarseness. We presented a patient with laryngeal granulocytic sarcoma involving the epiglottis, left vocal cord and aryepiglottic fold.

#### *Keywords*

*Larynx; granulocytic sarcoma; chronic granulocytic leukemia*

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### ÖZET

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Granülositik sarkom miyeloid lösemik hücrelerin ekstramedüller olarak depolanmış halidir. Bu tümörler akut miyeloid lösemisinin veya kronik miyeloid lösemisinin blast krizinin başlangıç döneminin spesifik lezyonları olarak kabul edilir. Granülositik sarkom vücudun herhangi bir yerinde oluşabilir. Hemopoetik tümörler tarafından larenksin tutulumu nadir bir olay olarak kabul edilmektedir. En sık görülen semptomlar ses kısıklığı ve nefes almada zorluktur. Biz burada epiglotu, sol vokal kordu ve ariepiglottik foldu tutan lareneal granülositik sarkomlu bir hastayı sunduk.

#### *Anahtar Sözcükler*

*Sarkoma; miyeloid; larinks tümörleri*

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Correspondence

**Mehmet YILMAZ, MD**

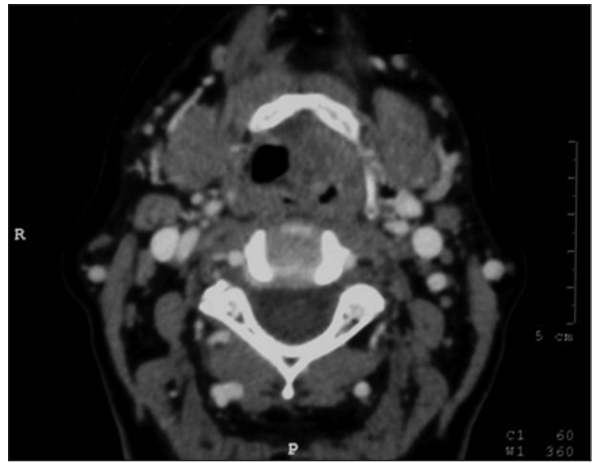
İstanbul University Cerrahpaşa Medical Faculty,  
Department of Otorhinolaryngology Head and Neck Surgery, İstanbul, Turkey  
E-mail: drmofly@yahoo.com

## INTRODUCTION

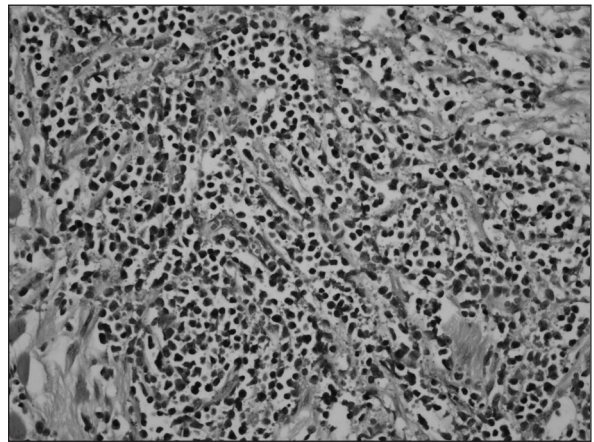
The granulocytic sarcoma (GS) is an extramedullary solid tumor composed of immature cells of granulocytic lineage (myelocytes).<sup>1</sup> The tumor may arise concurrently with chronic granulocytic leukemia (CGL), acute myeloid leukemia (AML), or myelodysplastic syndrome (MDS), or may precede the development of a disseminated leukemia.<sup>2</sup> The anatomic distribution commonly includes bones, nerves, lymph nodes and skin, but may involve a variety of soft tissues.<sup>3</sup> Laryngeal occurrence of GS is extremely rare. Surgery, radiation and chemotherapy are the treatment choices for GS.

## CASE REPORT

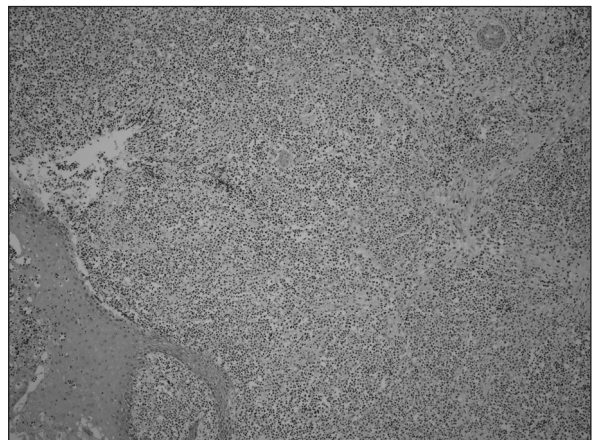
We present the case of 32 year-old-male, who was referred to our department with difficulty of breath and hoarseness. The patient had lost eight kilograms in the previous five months. The patient had no history of other systemic diseases. He had smoked one pack of cigarettes per day for ten years. Otolaryngologic examination with larynx endoscopy yielded an ulcerated and necrotising tumor on left side of laryngeal surface of epiglottis, on left vocal cord and left aryepiglottic fold. The left vocal cord was immobile. There were no masses in the neck. Computed tomography scan revealed a 2.5 cm × 3.0 cm mass at the laryngeal surface of epiglottis extending to left aryepiglottic fold (Figure 1). According to these findings and symptoms the patient was presumptively diagnosed with squamous cell larynx carcinoma. Multiple laryngeal biopsies were performed. Immunohistochemical examination of laryngeal specimen showed a dense, round to oval shaped, immature, hyperchromatic and cellular infiltrate with slightly polymorphic nuclei just beneath the epithelial layer (Figure 2, 3). With these findings the lesion was considered as granulocytic sarcoma. At the time of diagnosis there was no clinical or laboratory evidence of acute myeloid leukemia or chronic myeloproliferative disease. Four months after the diagnosis, bone marrow morphology was consistent with chronic myeloid leukemia, accelerated phase. He was referred to hematology clinic for chemotherapy treatment. The patient died approximately two months after the beginning of chemotherapy treatment.



**Figure 1.** Computed tomography scan revealed a 2.5 cm × 3.0 cm mass at the laryngeal surface of epiglottis extending to left aryepiglottic fold.



**Figure 2.** Larger magnification of the lesion demonstrating, round to oval shaped, immature, hyperchromatic and slightly polymorphic nucleus (H&E,400x).



**Figure 3.** Positive immunohistochemical staining of the tumor cells with myeloperoxidase (400x).

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## DISCUSSION

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Granulocytic sarcoma (GS), formerly referred to as chloroma, is an uncommon extramedullary tumor composed of dense aggregates of immature myeloid precursor cells.<sup>4</sup> Burns reported the first case in 1811.<sup>5</sup> The association of granulocytic sarcoma with leukemia was first made in 1892 by Dock.<sup>6</sup> It has most frequently (2.9-8%) been reported with the monocytic form of acute myelogenous leukemia (AML M5) but association with chronic myelogenous leukemia (3.9%) and at the onset of a blast crisis in chronic myelogenous leukemia is reported (4.2%).<sup>7</sup> In our patient GS has occurred in association with presence of chronic granulocytic leukemia. The frequency is higher among young patients, with a range of 2-81 years. There is a slight male predominance. In approximately 25% of cases, GS precedes the diagnosis of the underlying malignancy.<sup>2</sup> GS can appear in various sites including the skin, gingiva, lymph nodes, bone, soft tissue, and visceral organs and the tumor is capable of local tissue destruction and invasion.<sup>3</sup> GS rarely involves the larynx. Most common primary hemopoietic tumors of the larynx are extramedullary plasmacytoma and non-Hodgkin's lymphoma. Primary Hodgkin's disease, granulocytic sarcoma and mast cell sarcoma are extremely rare at this site. The prognosis of patients with laryngeal involvement in acute or chronic myeloid leukemia is always poor.<sup>8</sup> In our patient GS has involved laryngeal surface of epiglottis, left vocal cord and left aryepiglottic fold and local destruction and invasion sings have existed on laryngeal endoscopic view. The diagnosis of GS may be difficult. This is especially true if lesions are found prior to the diagnosis of hematologic malignancy, as in our patient. The differential diagnosis for granulocytic sarcoma includes malignant lymphoma, Ewing sarcoma,

inflammation, and giant cell granuloma. Misdiagnosis occurs at a frequency of 47% to 75%.<sup>9</sup> Pathological confirmation is important and in most cases is the final diagnostic clue. The pathological diagnosis of granulocytic sarcoma should be made on both histological and (immuno)histochemical grounds. The presence of eosinophils or other granulocytes should raise a suspicion for GS; however, special stains such as MPO, chloroacetate esterase, lysozyme, and immunophenotype are essential.<sup>10</sup> Recognition of this rare entity is important, because it saves the patient from an unnecessary operation, and aggressive induction chemotherapy can induce complete remission and improve the prognosis. Patients are routinely treated with chemotherapy, with or without radiotherapy, but as many as 85% relapse within 1 year.<sup>11</sup> Therefore, the definitive treatment should be chemotherapy followed by hematopoietic stem cell transplantation. When transplantation is not possible, we believe that chemotherapy might still improve survival rates if it is administered in higher doses than usual.<sup>12</sup> The mean interval between tumor diagnosis and the onset of over leukemia was reported to be 10 months, with a range of 1-49 months.<sup>2</sup> For the patients with AML mean interval is 4.4 months and 3.8 months for the patients with chronic granulocytic leukemia.<sup>7</sup> Our patient has died two months after the initial diagnosis despite of high dose chemotherapy treatment.

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## CONCLUSION

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The clinician must be aware of this rare condition. Primary tumors of the larynx must be clearly distinguished from laryngeal involvement by systemic or leukemic infiltrations. Surgery is not a treatment option for hemopoietic tumors. The treatment of granulocytic sarcoma must include radiation and aggressive chemotherapy.

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