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Unexpected Sequela of a Rapidly Growing Neck Mass: Histiocytic Sarcoma Resulting in Spontaneous Tumor Lysis Syndrome

Omar Z. Maniya[1*], Elizabeth Guardiani[2], Stanley H. Chia[3,4]
[1]Georgetown University School of Medicine, 3900 Reservoir Road, NW, Washington, DC 20057, USA
[2]University of Maryland School of Medicine, Department of Otorhinolaryngology, 655 W. Baltimore Street, Baltimore, MD 21201, USA
[3]MedStar Georgetown University Hospital, Department of Otolaryngology, 3800 Reservoir Road, NW, Gorman Building, First Floor, Washington, DC 20007, USA
[4]MedStar Washington Hospital Center, Department of Otolaryngology, 110 Irving Street, NW, GA-4, Washington, DC 20010, USA

***Corresponding Author:**
Name: Omar Z. Maniya
Email: ozm3@georgetown.edu

Abstract: Although neck masses are a common presenting symptom to an otolaryngologist, we report a rare case of a 27-year-old Hispanic female who presented with a six-week history of odynophagia, fevers and a rapidly enlarging right neck mass. Fine needle aspiration revealed large histiocytes and other inflammatory cells, but no evidence of malignancy. The patient was admitted to the hospital for hydration, parenteral antibiotics and scheduled for an open biopsy. Computed tomography at admission revealed a 6x6 cm neck mass, innumerable lung nodules, and ill-defined parenchymal lesions throughout the liver, spleen, and kidneys. Soon after admission, the patient had rapid clinical deterioration with profound lactic acidosis and multiple electrolyte abnormalities, and expired just two days after admission. Her final pathology was consistent with histiocytic sarcoma and cause of death was determined to be tumor lysis syndrome resulting from massive tumor burden.
Keywords: Otolaryngology, Neck Mass, Head and Neck Cancer, Histiocytic Sarcoma, Tumor Lysis Syndrome.

INTRODUCTION

Neck masses are a common reason for presentation to an otolaryngologist. However, otolaryngologists must be aware of potentially aggressive disease processes that are less frequently encountered. Histiocytic sarcoma (HS), a rare neoplasm of hematopoietic origin, presents at a median age of 46, with a slight male predilection. Approximately 1/3 of cases present in lymph nodes, 1/3 in skin and 1/3 in extranodal sites, with the GI tract being most common [1].

Tumor lysis syndrome (TLS) is a phenomenon that is well known to medical oncologists, but is less familiar to the otolaryngologist. Occurring primarily in hematopoietic tumors with a high proliferative rate and large tumor burden, TLS is most commonly associated with the initiation of cytotoxic therapy [2]. However, it is also known to occur spontaneously, particularly in hematopoietic neoplasms.

We describe a rare presentation of a histiocytic sarcoma neck mass that rapidly progressed to spontaneous tumor lysis syndrome.

CASE REPORT

A 27-year-old nonsmoking Hispanic female presented to the emergency department with a six-week history of odynophagia, fevers, and an enlarging right neck mass (Fig. 1). The patient had previously been treated with a course of antibiotics without improvement. CT with contrast revealed a 4 x 4 cm necrotic neck mass as well as a pulmonary nodule. The patient underwent a fine needle biopsy that revealed large histiocytes. She was admitted to the hospital one week later for dehydration and IV antibiotics. Repeat CT of the neck and chest revealed that the mass had increased in size to 6 x 6 cm (Fig. 2).

Biopsy at that time revealed sheets of large atypical cells with abundant foamy eosinophilic cytoplasm, large irregular nuclei, and prominent nucleoli. Staining was positive for CD4, CD68 and CD45, and negative for B and T cell markers as well as keratin and HMB-45.

The following day, the patient developed tachycardia, tachypnea, and abdominal distention. CT

angiogram showed no evidence of a pulmonary embolism and chest and abdominal CT scans revealed diffuse ascites with thickening of the bowel, as well as innumerable lung, liver, spleen, and kidney lesions (Fig. 3). An exploratory laparotomy revealed massive ascites with diffuse intra-abdominal tumor burden.

The next day, the patient developed spontaneous tumor lysis syndrome and became profoundly acidotic with significant hyperkalemia, hyperphosphatemia, and hypocalcemia despite aggressive correction. She continued to rapidly deteriorate; developing disseminated intravascular coagulation, multi-system organ failure, and decreased neurologic function. The patient died early the following morning, just two days after admission. Final pathology from the biopsy was conclusive for histiocytic sarcoma, and the patient's family denied a request for an autopsy.

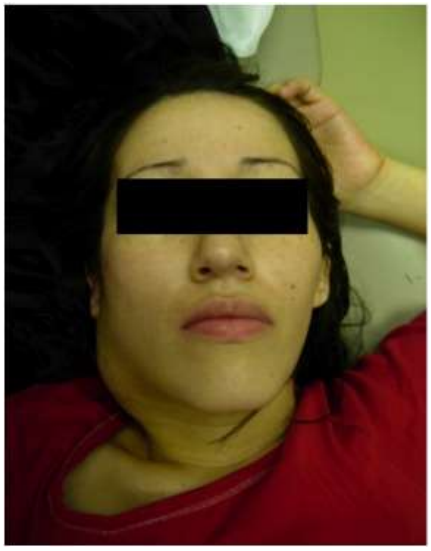

Fig. 1: 27 year-old non-smoking Hispanic female presents with an enlarging right neck mass

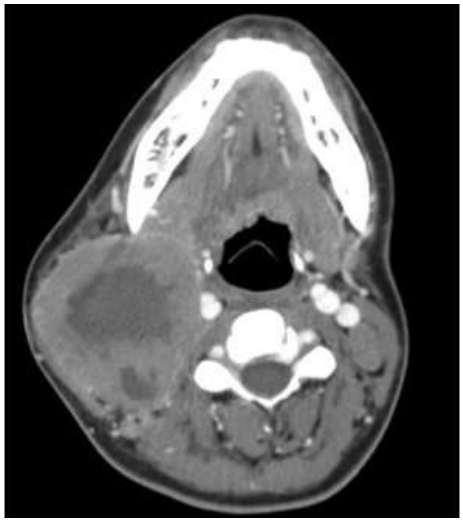

Fig. 2: CT of the neck with IV contrast shows a 6cm x 6cm necrotic neck mass

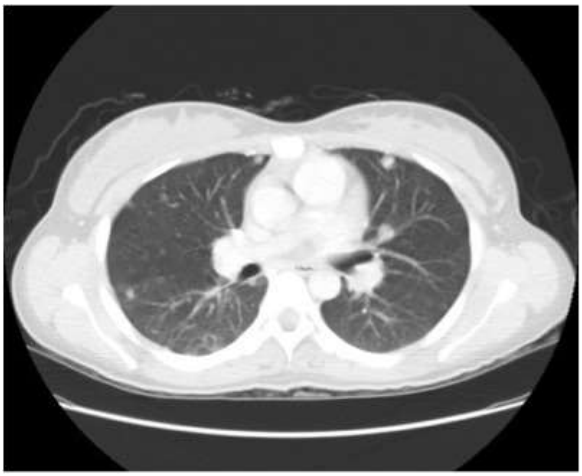

Fig. 3: Chest CT demonstrates multiple new pulmonary lesions consistent with metastases

DISCUSSION

Histiocytic sarcoma (HS) is a rare aggressive neoplasm of hematopoietic origin that has morphologic and immunophenotypic features that resemble mature tissue histiocytes. Diagnosis is made primarily by verification of histiocytic lineage and exclusion of other poorly differentiated large cell malignancies. This is done primarily by immunohistochemistry. Histiocytic markers such as CD68 and C163 are positive and B-cell, T-cell, epithelial, and melanotic markers are negative [1, 3, 4]. CD45 (common leukocyte antigen) is also generally positive [1].

The clinical course of HS is characterized by a rapidly progressive course and high mortality. Treatment of histiocytic sarcoma consists of wide local excision when possible with adjuvant radiotherapy [3]. Chemotherapeutic regimens have been proposed without evidence-based data [4]. Patients have a very poor prognosis with poor response to therapy except in cases of very limited disease where complete excision is possible [3, 4]. Our case is unique in that HS rarely presents as a neck mass, and therefore does not typically enter into the differential diagnosis of otolaryngologists. However, HS has previously been described in the thyroid gland, the mandibular condyle and the palate [3-5].

The exact mechanism of spontaneous tumor lysis syndrome (TLS) is not well defined, but is thought to be related to rapid tumor necrosis. Patients with bulky lymphadenopathy, elevated LDH, elevated WBC counts, pre-existing renal disease and hyperuricemia are at increased risk of TLS [6]. This results in the release of intracellular ions with subsequent hyperkalemia, hyperphosphatemia, hyperuricemia and hypocalcemia

secondary to calcium phosphate precipitation. This leads to metabolic acidosis and acute kidney injury [2, 5]. Tumor lysis syndrome can be classified as either laboratory or clinical. Laboratory TLS is defined as 2 or more of the metabolic abnormalities (hyperkalemia, hyperphosphatemia, hyperuricemia or hypocalcemia) occurring within the same 24-hour period. Clinical TLS is diagnosed when laboratory TLS occurs in conjunction with acute kidney injury, seizures, cardiac dysrhythmias or death [2, 7].

Treatment must be initiated promptly and typically consists of IV hydration, urine alkalinization, correction of electrolyte abnormalities, and hypouricemic agents. Occasionally, loop diuretics or hemodialysis are employed to treat life-threatening electrolyte abnormalities or volume overload. Of the hypouricemic agents used, administration of rasburicase (recombinant urate oxidase) can be useful to improve renal function and lower phosphorus levels, and may be more effective than allopurinol (xanthine oxidase inhibitor) in preventing tumor lysis syndrome [2, 7]. Rasburicase is recommended as first line treatment in patients who are at high risk for developing clinical tumor lysis syndrome [8]. However, cost considerations limit the use of rasburicase in patients with low to intermediate risk of developing TLS [7]. Tumor lysis syndrome is considered a medical emergency and is associated with a high rate of renal failure and death [2].

CONCLUSION

Although neck masses are a common reason for presentation to an otolaryngologist, tumor lysis syndrome is less frequently encountered. This aggressive disease process must be diagnosed and treated quickly if patients are to have any reasonable chance of recovery.

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