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CLINICAL VIGNETTE

Chronic sinusitis: A rare presentation of Acute Myeloid Leukemia

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A 66-year-old woman with a past medical history of hypertension and asthma presented for a second opinion regarding 8 months of ongoing sinus congestion. She denied fevers and chills but had a cough productive of thick mucus and poor upper airway air movement. She had sought care through her primary care physician and at least two different Ear, Nose and Throat (ENT) specialists and had completed several rounds of steroids, and balloon septoplasty without improvement in her symptoms. She subsequently developed bilateral hearing loss, which led to bilateral tympanostomy tube placement. The patient denied cultures or biopsies having been taken up to that point; and denied receiving antibiotics. She was noted to have a pet dog and while it had been suggested that her symptoms might be related to pet dander allergy, she was unable to avoid contact with the pet. In an attempt to avoid additional surgery, she sought out a second opinion.

On physical exam she was alert and fully oriented. Vital signs were within normal limits. Audible sinus congestion was apparent, with essentially absent air movement through the nasal passages bilaterally. The distal nasal turbinates were free of erythema, while the visualized more proximal turbinates were boggy and indurated bilaterally, without significant discharge or ulceration. The oropharynx was clear of adenopathy, exudates, or erythema. Shotty adenopathy was noted in the anterior and posterior cervical chains of the neck. She had diminished hearing bilaterally despite clear external auditory canals and well healed bilateral tympanostomy tubes. Chest was remarkable for bibasilar crackles, and a 1/6 flow murmur was auscultated at the right and left upper sternal borders. Abdominal, neurological, musculoskeletal, skin, and psychiatric examinations were normal.

A chest x-ray was without acute process. The patient was referred to a new ENT specialist for further evaluation and labs were collected.

Labs and Studies

Initial lab results demonstrated a normal complete metabolic panel and lactate dehydrogenase (222U/L). A complete blood count revealed leukocytes 4 x10E3/uL, hemoglobin (Hg)14g/dL, hematocrit 42%, platelets 239 x10E3/uL. White blood cell count differential revealed low absolute neutrophil count at 756 cells/uL, normal lymphocytes at 2676 cells/uL, low monocytes at 92 cells/uL, with normal eosinophils and basophils at 444 and 32 cells/uL respectively.

Initial Treatment Course

The patient was evaluated by ENT within two weeks and was scheduled for polypectomy ten days thereafter. Results of tissue and bone biopsy collected intraoperatively were consistent with granulocytic sarcoma (GS).

The patient was sent to Oncology for further evaluation and underwent marrow aspirate with abnormal DDIMER > 10,000 and fibrinogen 70 mg/dL consistent with disseminated intravascular coagulation (DIC). In addition, her total white count had dropped to 2.96 x10E3/uL, Hg to 8.1g/dL, platelets to 154x10E3/uL, and a manual differential identified 17% circulatory blasts. The patient was admitted for DIC and her marrow aspirate confirmed acute myeloid leukemia (AML).

The patient was started on induction therapy with cytarabine and idarubicin 7+2, and day 14 bone marrow aspiration revealed 10% blasts. She underwent re-induction with 5+2 after which she achieved full remission. She is now residing with her adult daughter; together they are scheduled for allogenic stem cell transplant.

Discussion

Granulocytic sarcomas are extramedullary proliferations of cells stemming from the granulocyte series[1,2]. Burns first described the tumor in 1811[3] when it was identified in a patient's orbit; in 1853,

the term *chloroma* appeared[4], referencing the green color of the tumors due to the myeloperoxidase contained within them[3]. In 1966, the more appropriate term, *granulocytic sarcoma* (owing to the tumor's mesodermal origin), was coined[3]. Alternatively, the term *myeloid sarcoma* has also been used to describe the tumor.

Granulocytic sarcomas typically exist in combination with AML and they were first associated with acute myeloid leukemia in 1893[5]. In 1981, this association was further characterized by Neiman et al. in a study of 61 cases of granulocytic sarcoma[6]. The most common sites of granulocytic sarcoma occurrence were in the bone/periosteum, soft tissue, lymph nodes, and skin; but could arise in essentially any tissue. Not all patients with granulocytic sarcoma develop AML; granulocytic sarcomas are now known to be associated with other myeloproliferative diseases as well[7].

Our patient's case is uncommon in several ways; first granulocytic sarcomas are rare regardless of location; being observed in only 3-9.1% of AML cases[3]. In one case series of hematolymphoid malignancies of the head and neck, it was found that of 122 malignancies, only 1 was leukemoid in nature; the other 121 were lymphoid in origin[8]. Second, GS rarely occurs in adults; it typically appears in patients under the age of 15[9]. Third, GS is a highly atypical primary presentation of AML or preexisting pancytopenia. In fact, less than 1% of patients with AML present in this manner[10]. Finally, our patient's GS was identified in the nasal cavity. Granulocytic sarcoma occurrence is well documented in the head and neck; however, it has a predilection for the oral cavity, as opposed to the sinuses[11]. We found only one other case of an *initial presentation* of AML characterized by GS in the sinus cavity[7]. Two other cases of nasal cavity GS were identified, however they were in patients with existing diagnoses of AML[12,13].

Alternatively, our case is relatively "classic" in that of patients identified to have a primary presentation of GS, a diagnosis of acute leukemia typically follows in the following 2 years time (average 10 months)[6]. Our patient was found to have AML eight months after symptom onset; and within one month of initial evaluation.

Acute myeloid leukemia should always be a part of the differential when granulocytic sarcomas are identified. In cases that have an initial presentation of GS as opposed to the hallmark findings of AML such as pancytopenia, it can delay diagnosis, as was seen in our patient. Generally, granulocytic sarcomas are poor prognostic signs in patients with AML[1], and it has been estimated that life expectancy in patients with AML in association with a granulocytic sarcoma is only eight months[14]. Granulocytic sarcomas are treated by treating the underlying AML with standard dose cytarabine and an anthracycline. For those sarcomas that cause mass effect, radiation therapy can also be considered[15].

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