Review Article

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Chronic Myelomonocytic Leukaemia in a Man with Acute Renal Damage

Hetty Carraway*

Department of Hematology and Bone Marrow Transplantation, Rambam Health Care Campus, Haifa, Israel

*Author for correspondence: hetty@Carraway.health.gov.il

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Abstract

Myeloproliferative disease (MPD) and the myelodysplastic syndrome share characteristics with chronic myelomonocytic leukaemia (CMML), a relatively uncommon clonal hematologic condition. Rarely is extra medullary leukemic involvement a CMML presenting characteristic. The therapy of patients with extra medullary symptoms is difficult because there are no precise standards for treatment. In this report, we go through how our patient, who had CMML and submandibular lymphadenopathy and gingivitis when they first appeared, was treated.

Introduction

The myelodysplastic syndrome and myeloproliferative neoplasms (MDS/MPN) share clinical and pathological characteristics with the malignant hematopoietic stem cell condition known as CMML. CMML has a median survival of 14 to 22 months and a median age at diagnosis of 71 years. Extra medullary symptoms of CMML, which can affect the spleen, liver, skin, and lymph nodes, are rare. Here, we present a case of CMML with gingival infiltration and extra medullary symptoms of myeloid sarcoma (MS) of the lymph node.

Acute pulmonary problems in patients with children acute myelogenous leukaemia (AML) enhance the chance of early death. The plausible aetiologies have been identified as pulmonary leukostasis and systemic inflammatory response syndrome (SIRS) following leukaemia cell lysis. In this case study, we describe an 18-month-old girl who developed cardiopulmonary failure and needed ECMO support after starting chemotherapy for acute myelomonocytic (M4eo) leukaemia [1].

Haematology, oncology, rheumatology, and virology all cross in the rare polyclonal B lymph proliferative condition known as MCD. Multiple organ system dysfunctions, reactive proliferation of morphologically benign cells, and recurrent systemic inflammatory symptoms are all signs of MCD, which has a diverse range of aetiologies. Myelodysplastic syndromes (MDSs) and myeloproliferative neoplasms (MPNs) share characteristics with chronic myelomonocytic leukaemia (CMML), a clonal stem cell illness with a mixed but generally poor prognosis. With a range of 65 to 75 years for the median age upon diagnosis, it nearly exclusively affects the elderly. The pathophysiology of CMML appears to be complex, as is the case with MDS, and is connected to altered genetic, molecular, micro environmental and immunologic states in various individuals [2].

Compared to healthy controls, patients with MCD have a noticeably higher chance of developing severe pancytopenia, multiorgan failure, and lymphoma. On the other hand, the progression of MCD into CMML, which is associated with many pathogenic cell types, is relatively unusual. In this instance, we will first go through an MCD patient who developed CMML and responded favourably to decitabine medication. Myeloid sarcomas are tumour masses that develop in extra-medullary sites throughout the body and are made up of myeloid blasts or immature myeloid cells. Although the skin, lymph nodes, bone, soft tissues, and gastrointestinal tract are the most often affected areas, these neoplasms can occur anywhere and are frequently accompanied by bone marrow involvement of an underlying hematologic malignancy. Instead of having a typical presentation, myeloid sarcomas can have a variety of clinical symptoms that rely on the size and location of the

tumour as well as the effects of underlying hematologic conditions (such as infection, haemorrhage, and organomegaly) [3].

The histomorphologic diagnosis of myeloid sarcomas is usually challenging or delayed in the absence of a prior diagnosis of a blood-born malignancy since they are less common than many other solid tumours. Myeloid sarcomas are sometimes initially confused with aggressive big B-cell lymphomas due to their histomorphologic similarities to non-Hodgkin's lymphomas. But it's important to get the right diagnosis because the chemotherapeutic regimens for treating acute non-lymphoblastic leukaemias and malignant lymph proliferative diseases are very different from one another. Unexpectedly, neither the clinical characteristics nor the therapeutic responses of myeloid sarcomas seem to be affected by the patient's age, gender, anatomic location, personal association with acute myeloid leukaemia (AML), myelodysplastic syndrome (MDS), or myeloproliferative disorders (MPD), histomorphologic sub classification, or the vast majority of cytogenetic findings [4].

In the largest series of adult-derived myeloid sarcomas to be described to date, tumours were discovered to develop most commonly in the presence of prior or contemporary AML (41%), as well as in cases of de novo neoplasms (27%), and less frequently in the presence of prior or concurrent MPD (15%) or MDS (15%). In contrast to cases linked to MPD, MDS, or chronic myelomonocytic leukaemia (CMML), myeloid sarcomas of the testis exhibit a more pronounced connection with AML (68%), which has been described only infrequently. In this report, we present a rare instance of testicular myeloid sarcoma, where the early clinical symptoms were refractory pericarditis and a unique pattern of migratory arthralgia's and myalgia's limited to the upper extremities. The patient's initial symptoms were attributed to CMML-related autoimmunity because of the patient's rapid and drastic improvement after therapy, which led to the concomitant diagnoses of myeloid sarcoma and CMML. A brief summary of this unusual case is given below, along with a discussion of how certain underlying hematologic conditions linked to myeloid sarcomas, specifically MDS and CMML, can expand the range of presenting symptoms related to these neoplasms because of their propensity to cause autoimmune rheumatologic phenomena [5].

Discussion

The order in which the symptoms first manifested it is the most striking aspect of the instance discussed above. Myeloid sarcomas are solid tumours, and they frequently manifest early on or present with symptoms of mass effect. But in this case, acute pericarditis and arthralgia's/myalgia's of the upper extremities were the initial symptoms that made the patient go to the doctor. When these early symptoms first appeared, about 3 months before the first instance of scrotal asymmetry, there was no obvious sign of an underlying hematologic malignancy. Furthermore, after radical orchiectomy and the start of induction chemotherapy, both of which were connected to the immediate bulk removal of antigenic cancer cells, these symptoms were quickly alleviated, albeit momentarily [6]. According to this research, the paraneoplastic manifestations of the concurrently diagnosed CMML are the symptoms existing before the initial identification of testicular enlargement. The clinical manifestations of paraneoplastic autoimmune phenomena include acute systemic vasculitis disease, isolated chronic autoimmune phenomena, classical connective tissue disorders, immune-mediated cytopenia, and abnormal serum immunologic results. They are seen in about 10% of patients with MDS and CMML. Because of this, we had a strong suspicion that the patient's pleuritic chest discomfort, arthralgia, and myalgia's, as well as the IgM-aCL hypercoagulable state that subsequently exacerbated his clinical course, were likely caused by autoimmunity associated with his underlying CMML [7].

In the current instance, our patient experienced aortitis brought on by CMML. A prominent distinguishing characteristic of CMML, a clonal disease of a bone marrow stem cell, is monocytosis. It was initially categorised as a wholly myelodysplastic condition. Clinicians now understand that CMML has both dysplastic and proliferative characteristics. The WHO categorises CMML as a myelodysplastic/myeloproliferative disease (MDS/MPD) as a result of these two traits. SIADs (systemic inflammatory and autoimmune disorders) are found in 10–20% of MDS patients [8].

Vasculitis, a rare but well-recognized paraneoplastic consequence of MDS and MPD diseases, primarily affects small and medium blood arteries and very infrequently larger

ones like the aorta. Vasculitis that is associated with MDS/MPD typically manifest as skin lesions, arthralgias, and neuropathy. The two conditions most frequently linked to MDS are polyarthritis nodosa and leukocytoclastic vasculitis. Only three case reports describing aortitis especially worsening CMML were found in our literature search. MDS/MPD syndromes have been associated with immunological abnormalities, which include both humoral and cellular abnormalities. Although the precise underlying mechanisms are yet unknown, it has been speculated that dysregulated antigenic presentation with on-going immunological stimulation, inadequacies in lymphocyte function, flawed natural killer cell function, flawed phagocytic cell function, and abnormalities in lymphocyte function may all be involved.

Steroid medication virtually instantly reduced the acute systemic inflammatory signs in our patient. However, the patient's CMML advanced very quickly, and three months after her aortitis diagnosis, she passed away from leukemia-related complications. In a multicenter retrospective study conducted in France, the diagnosis of SIADs associated with MDS occurred before the diagnosis of MDS in 37% of patients, concurrently in 31% of cases, and after the diagnosis of MDS in 32% of cases, with an average interval of 8.6 months between the two diagnoses [9].

Corticosteroids and a targeted approach to treating MDS/MPD symptoms make up the initial vasculitis treatment. One study of 26 individuals with CMML worsened by SIADs found that 87% of them responded quickly to steroids alone. Nevertheless, 40% of patients required a second-line treatment because to recurrence or steroid dependency despite this promising first response. Similar outcomes for MDS-associated SIADs were found in a bigger research, where 80% of patients responded to corticosteroids alone and 50% needed a second-line therapy for the same reasons as mentioned above. Most cases of CMML and other MDS patients who received treatment with the hypo ethylating drugs azacitidine and decitabine exhibited an improved response, with much lower levels and steroid dependence rates [10].

Conclusion

This example of primary adrenal insufficiency caused by prolonged posaconazole use is presented. Clinicians need to be aware of this potential long-term side effect as posaconazole use grows for treating and preventing invasive fungal infections. It should not be administered to individuals who are at danger of developing adrenal insufficiency, according to clinicians. Weight loss, hypotension, or hypoglycemia, which is clinical indications or symptoms of adrenal insufficiency, may be red flags that this side effect is developing. Isavuconazole, the newest azole to receive approval, may also call for increased awareness of this side effect.

Conflicts of Interest

None

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