Tennessee Medicine E-Journal

Volume 2 | Issue 2 Article 5

August 2016

A Case Of Relapsing Polychondritis In A Patient With Acute Myeloid Leukemia With Myelodysplasia Related Changes

Tugcan Alp
Uludag University, Bursa, Turkey, tugcanalp82@hotmail.com

Hilmi Erdem Gozden
Uludag University, Bursa, sermin10@yahoo.com

Tuba Ersal
Uludag University, Bursa, tubaersal@hotmail.com

Vildan Ozkocaman
Uludag University, Bursa, vildanoz@uludag.edu.tr

Fahir Ozkalemkas
Uludag University, Bursa, fahir@uludag.edu.tr

See next page for additional authors

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Authors
Tugcan Alp, Hilmi Erdem Gozden, Tuba Ersal, Vildan Ozkocaman, Fahir Ozkalemkas, and Rıdvan Ali

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INTRODUCTION

Relapsing polychondritis (RP) is an uncommon multi systemic disease most commonly presenting as recurrent inflammation of the cartilage of the ears, nose, joints, respiratory tract and non-cartilaginous tissues such as kidneys, blood vessels [1,2,3]. The pathogenesis of the disease is not clear but the frequent presence of antibodies to type II collagen which gives the possibility of an autoimmune background [1]. The diagnosis is based on the clinical symptoms. Relapsing polychondritis is associated with malignancies, myelodisplastic syndrome (MDS), solid tumors, rheumatologic or autoimmune diseases [1,4].

Myelodysplasia is a term used to encompass a spectrum of clonal myeloid disorders marked by ineffective hematopoiesis cytopenias, qualitative disorders of blood cells and their precursors, clonal chromosomal abnormalities and a variable predilection to undergo clonal evolution to polyblastic acute myelogenous leukemia (AML). The disorders range from relatively indolent clonally derived anemias, with a relatively lower frequency of progression to AML, to more troublesome clonal multi lineage cytopenias or to oligoblastic myelogenous leukemias that often progress to overt AML [5].

CASE

A 58-year-old male patient presented to the gastroenterology department, complaining of weakness, paleness, nausea and vomiting. There were no unusual features in patient's history and family history. On his examination; paleness, hepatomegaly were found, the spleen was not palpable, but Traube's space dullness was determined. Gastritis was established by upper gastrointestinal endoscopy. The Laboratory tests revealed leukocytosis (30.900/mm^3), anemia (6.18 g/dL) and thrombocytopenia (114.000/mm^3). Hence, the patient was referred to hematology department for further investigation. In the peripheral blood smear; there were 50% blasts, %1 atypical lymphocyte, and %15 erythroblasts. He was applied bone marrow aspiration and revealed hypercellularity, myeloblastic infiltration (35%), megaloblastic and dysplastic erythroblasts significantly. According to these findings, acute myeloid leukemia with myelodysplasia-related changes was diagnosed. So, the patient was hospitalized and started to receive remission induction chemotherapy protocol, idarubicin and cytosine arabinoside. No cytogenetic anomaly related acute leukemia existed by fluorescence in situ hybridization and polymerase chain reaction. His chromosomal analysis was 46XY. The sixth day of the chemotherapy the patient had a fever and we started piperacilin+tazobactam empirically after taking blood and urine culture samples. We changed antibiotics to meropenem and vancomycin because of ongoing fever. We noticed swelling, rubescence and pain on the right pinna and next day on the left pinna on daily examination (Figure 1). He also complained of pain, rubescence and swelling on the left periorbital area and hoarseness. With this presentation, we considered a complicated soft tissue infection and added levofloxacin, sulbactam and daptomycin instead of vancomycin. The patient consulted with otorhinolaryngologist and an ophthalmologist. The otorhinolaryngologist diagnosed relapsing polychondritis on the second examination since the patient had inflammation on both of the pinnas and hoarseness. Orbital MRI was performed and evidence of inflammation was found. The ophthalmologist and otorhinolaryngologist suggested continuing the present therapy. On the following days, complaints about the ears and hoarseness reduced spontaneously. The blood and urine cultures were negative.

We stopped the antibiotics when the patient were afebrile for and had 1500/mm ^{3} neutrophile count. On the 32nd day of the chemotherapy, bone marrow aspiration was performed and remission was observed in the bone marrow aspiration. The patient who had remission and recovery was discharged.

DISCUSSION

Relapsing polychondritis is an uncommon inflammatory condition that first characterized by Jaksch-Wartenhorst in 1923 [6]. Diagnostic criteria were first described by McAdam et al. in 1976 (3 of 6 clinical features necessary for diagnosis); bilateral auricular chondritis, nonerosive seronegative inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, audiovestibular damage. Damiani and Levine introduced the second criteria in 1979 (1 of 3 conditions necessary for diagnosis); Three McAdam et al. criteria or one McAdam et al. criterion plus positive histology results or two McAdam et al. criteria plus therapeutic response to corticosteroid or dapsone therapy was present. The third criteria defined by Michet et al. in 1986 (1 of 2 conditions necessary for diagnosis); proven inflammation in 2 of 3 of the auricular, nasal, or laryngotracheal cartilages, proven inflammation in 1 of 3 of the auricular, nasal, or laryngotracheal cartilages plus 2 other signs including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis, and hearing loss [7,8,9]. The most common symptom is auricular chondritis and arteritis. As in the third criteria, our case had inflammation of bilateral auricular and laryngotracheal cartilages.

The association of MDS and autoimmune diseases are frequent. Relapsing polychondritis is an autoimmune phenomenon that was described 0.7-5.4% in MDS series [10]. It is considered that MDS may have a poor prognosis because of the occurrence with RP [11]. Myelodysplastic syndrome and RP may come in view concurrently, or one disease follows the other one, as in our case [2]. Our case was associated with myelodysplasia, subtype refractory anemia with excess blasts and acute myeloid leukemia transformation [12]. Toprak et al. reported a case, which was diagnosed RP during chemotherapy of acute myeloid leukemia [13]. Our case also had RP during remission induction chemotherapy process.

RP can easily be misdiagnosed as an infection because of relapsing course and response to antibiotics [14]. Corticosteroids are the major treatment. Nonsteroidal anti-inflammatory drugs, dapsone, colchicine, azathioprine, methotrexate, cyclophosphamide, hydroxychloroquine, cyclosporine, and infliximab are other effective therapies [1]. Surgical treatment may be necessary for respiratory, cardiovascular complications and nose deformity [15]. Clinical pattern varies from low-grade up to rapidly progressive disease. Besides, these treatment choices, spontaneous remissions are common, as in our case [1].

In this report, we wanted to notice a rare condition that the association of RP and AML with myelodysplasia-related changes and MDS and autoimmune diseases.

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