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Pancreatic involvement of acute lymphoblastic leukemia in an adult patient: A Case Report

Yetişkin bir hastada akut lenfoblastik löseminin pankreas tutulumu: Bir Olgu Sunumu

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ABSTRACT

Acute lymphoblastic leukemia, it is responsible for 80% of childhood leukemia and accounts for 20% of all adult leukemias. Granulocytic sarcoma is an extramedullary tumor arising from immature myeloid serial cells and is seen in 2.5-9.1% of acute leukemia patients. Although it is generally seen in the subperiostal region of bone and soft tissue, it can be detected in all parts of the body. Granulocytic sarcoma is frequently diagnosed in pediatric patients, although it affects both pediatric and adult patients (cases between 3 months and 89 years of age have been reported). The main treatment of granulocytic sarcoma is chemotherapy to achieve cytoreduction and remission. Chemotherapy consists of remission induction, central nervous system prophylaxis, and post-remission treatment. Radiotherapy is used in symptomatic granulocytic sarcoma, spinal cord compression, vena cava superior syndrome and respiratory tract compression due to mediastinal mass. We present a case in which we applied radiotherapy because of pancreatic involvement of acute lymphoblastic leukemia in an adult patient. We analyze through these observations the clinical, histological, and therapeutic characteristics of this entity.

Key Words: Acute lymphoblastic leukemia, Granulocytic sarcoma, Radiotherapy

ÖZET

Akut lenfoblastik lösemi, çocukluk çağı lösemisinin %80'inden sorumludur ve tüm yetişkin lösemilerin %20'sini oluşturur. Granülositik sarkom, immatür miyeloid seri hücrelerden kaynaklanan ekstramedüller bir tümördür ve akut lösemi hastalarının %2.5-9.1'inde görülür. Genellikle kemik ve yumuşak dokunun subperiostal bölgesinde görülmekle birlikte vücudun her yerinde tespit edilebilir. Granülositik sarkom hem pediyatrik hem de yetişkin hastaları etkilese de (3 ay ile 89 yaş arasında vakalar bildirilmiştir) pediatrik hastalarda sıklıkla teşhis edilir. Granülositik sarkomun ana tedavisi sitoredüksiyon ve remisyon sağlamak için kemoterapidir. Kemoterapi, remisyon indüksiyonu, merkezi sinir sistemi profilaksisi ve remisyon sonrası tedaviden oluşur. Radyoterapi semptomatik granülositik sarkom, omurilik kompresyonu, vena kava superior sendromu ve mediastinal kitleye bağlı solunum yolu kompresyonunda kullanılır. Erişkin bir hastada akut lenfoblastik löseminin pankreas tutulumu nedeniyle radyoterapi uyguladığımız bir olguyu sunuyoruz. Bu gözlemler aracılığıyla bu varlığın klinik, histolojik ve terapötik özelliklerini analiz ediyoruz.

Anahtar Kelimeler: Akut lenfoblastik lösemi, Granülositik sarkom, Radyoterapi

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INTRODUCTION

Acute lymphoblastic leukemia (ALL) is the most common neoplastic disease for children under 15 years of age and is responsible for 80% of childhood leukemia.[1,2] The incidence of the disease in adults is between 1-2/100,000 and it constitutes 20% of all adult leukemias.[3] General systemic findings are; fever (60%), weakness (50%), pallor (40%), bone marrow (normocytic) neutropenia due to occupational anemia, thrombocytopenia, lymphadenomegaly and hepatosplenomegaly.[4] Granulocytic sarcoma (GS) is an extramedullary tumor arising from immature myeloid serial cells and is seen in 2.5-9.1% of acute leukemia patients. Although it is generally seen in the subperiostal region of bone and soft tissue, it can be detected in all parts of the body.[5,6] The main treatment of GS is chemotherapy to achieve cytoreduction and remission. Chemotherapy consists of remission induction, central nervous system (CNS) prophylaxis, and post-remission treatment.[7] Radiotherapy is used for symptomatic chloromas, spinal cord compression, vena cava superior syndrome and respiratory tract compression due to mediastinal mass. Steroids and chemotherapy are also resolves these complications. Cranial radiotherapy is required for CNS involvement that does not heal. In this report, an extremely rare case of ALL with pancreatic involvement is discussed in the light of the literature.

CASE REPORT

A 35-year-old male patient was admitted on December 19, 2015 with a 3-week-long back pain, leg pain and widespread ecchymoses on the body. On physical examination paleness and ecchymosis of the skin was observed. Laboratory findings were showing signs of, anemia and elevated liver function. Hepatosplenomegaly was seen in abdominal ultrasonography and computed tomography. The bone marrow biopsy analysis showed hypercellular bone marrow with acute leukemia infiltration. In the bone marrow aspirate, almost completely narrow granule-free basophilic cytoplasm and 1-2 nucleolus blastic cells were seen. Atypical large cells were positive for CD10, CD19, CD20, HLA-DR in flow cytometric analysis and immunohistochemical study. In a molecular cytogenetic analysis study t (9; 22) (q34; q11.2) BCR/ABL1 was 85% positive. The patient was diagnosed with B-cell ALL. PETHEMA LPA-99 protocol was applied to the patient on 22.12.2015, 02.03.2016 and 21.04.2016. Allogenic bone marrow transplantation was made on 22.12.2016. In one year follow up, patient had admitted to hospital for abdominal pain and a mass with 36x38 mm diameter in the head and neck of the pancreas was detected in abdominal MRI scans (Figure 1) (19.11.2017). Fine needle aspiration biopsy of pancreas revealed leading Precursor B cell lymphoblastic leukemia infiltration (Figure 2). The patient received 24Gy (2Gyx12 fraction) radiotheraphy to the pancreas 07.12.2017. Cheomotherapy was given to the patient whose pain palliaation was completed.

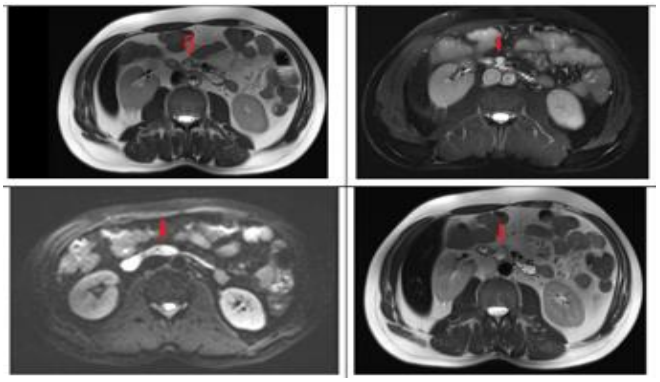

Figure 1. MRI image of the pancreas

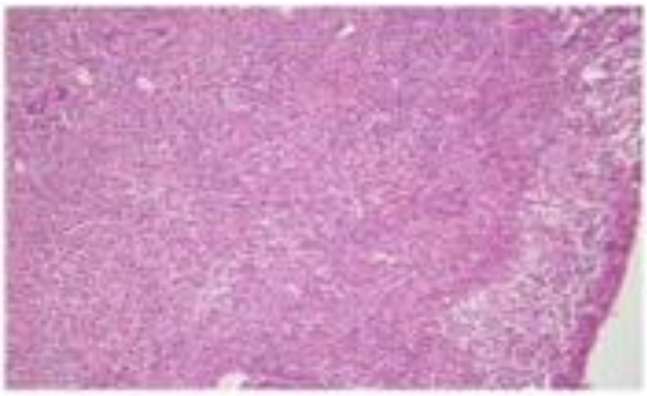

Figure 2. Photomicrograph of specimen shows immature and mature myeloid cells (H and E, ×100). The diagnosis was confirmed as myeloid sarcoma according to immunohistochemical staining.

DISCUSSION

Extramedullary involvement of ALL, is a rare and clinically important condition, arising from immature myeloid serial cells in leukemia patients is defined as chloromas or granulocytic sarcoma.[7-9] Although it is usually seen in bone and soft tissue, it can be detected any where in the body.[10] Although GS affects both pediatric and adult patients (cases have been reported between 3 months and 89 years old), it is diagnosed

frequently in pediatric patients.[11,12] Our case was a 35-year-old male patient. Classically, patients with acute leukemia have clinical signs of paleness and fatigue caused by pancytopenia and anemia. However, patients with pancreatic involvement may experience symptoms of abdominal pain, jaundice, or acute pancreatitis. Some patients may present with nonspecific symptoms such as persistent or intermittent fever, similar to a viral infection. In our case, pancreatic involvement gave symptom with abdominal pain. ALL led to significantly improved results with the new protocols, and survival rates were approximately 70% and 90% in adults and children, respectively. Adequate CNS prophylaxis is and mediastinal irradiation contributed to therapeutic success.[13] It is recommended that leukemias should be treated with more aggressive chemotherapy protocols in patients with multiple extramedullary organ involvement, including pancreas.[14] Our patient first applied mass-directed radiotherapy. Chemotherapy was then given.

CONCLUSION

Pancreatic involvement of leukemia is an extremely rare but clinically important cause in patients with a history of leukemia. Abdominal imaging should be undertaken in patients with ALL having gastrointestinal system symptoms. But, the leukemia of the pancreas cannot be distinguished from pancreatic tumors based on imaging findings alone. Therefore, histopathological confirmation is mandatory. Because it is known that extramedullary involvement of leukemia responds to systemic antileukemia at a high rate. Thus, unnecessary, aggressive surgery can be avoided.

Authorship contribution statement

Consept and desing: MK
Acquisition of data: MK and YK
Analysis and interpretation of data: MK and YK
Drafting of the manuscript: MK
Critical revision of the manuscript for important intellectual content: MK
Statistical analysis: MK

Declaration of competing interest

None of the authors have potential conflicts of interest to be disclosed.

Ethical approval

Not applicable.

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