Paraspinal Masses Due to Myeloid Metaplasia

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Received Date: April 08, 2016, Accepted Date: May 13, 2016, Published Date: May 23, 2016.

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Abstract

We present the case of a 34-year-old male homozygous for sickle cell disease found to have asymptomatic bilateral paraspinal masses (PSM) on chest x-ray, further visualized on Chest Computer Tomography (CT) scan with intravenous (IV). The unusually large PSM were attributed to extramedullary hematopoiesis (EMH) owing to erythroid hyperplasia. Follow-up CT scan after five years showed no significant changes in the bilateral paravertebral masses consistent with the diagnosis of EMH.

Keywords: Extramedullary Hematopoiesis; Paraspinal Masses; Sickle Cell Disease

Introduction

PSM are infrequently encountered and the differential diagnosis is wide. Neoplastic processes are the most common culprits with particular predilection for metastatic adenocarcinoma [1]. Although less frequent, benign etiologies such as tuberculosis and EMH should be considered in the appropriate clinical settings [1,2]. We herein present a case of unusually large PSM attributed to EMH in a young patient with sickle cell disease.

Case Report

A 34-year-old African-American male with homozygous sickle cell disease presented to the hospital with shortness of breath and generalized bony and joints pain. He was diagnosed with sickle cell crisis, treated with IV (Intravenous) fluids, pain control and was subsequently transfused with two units of packed red blood cells. Chest X-ray on admission showed widening of superior mediastinum (Figure 1) with no infiltrates. CT scan of chest with IV contrast showed two large retrocardiac PSM with the left mass measuring 6.8 cm/4.2 cm and the right mass measuring 5.9 cm/3.7 cm (Figure 2). The masses were well-marginated, showed no evidence of erosions onto the adjacent vertebral bodies, no widening of the neural foramina was seen and no enlarged intrathoracic lymph nodes were noted. Clinically, the patient improved following treatment, and no symptoms could be attributed to the presence of the bilateral PSM. A careful and detailed history was taken to further narrow the differential diagnoses of these masses. The absence of involvement of adjacent structures such as bones and the lack of mediastinal inflammation along with the lack of adenopathies made infectious etiologies unlikely. Furthermore the interferon-gamma release assays (quantiferon) for latent tuberculosis (TB) was negative ruling out TB as a possible culprit of PSM [3,4]. Another differential diagnosis to be considered is paraganglioma and multiple nerve sheaths tumors [2]. Despite the lack of histological diagnosis, the location of these tumors adjacent to the neural foramina and the presence of intrasional fat densities on imaging in a background of chronic anemia favored EMH. Intrasional fat is suggestive but not confirmatory of EMH since bone marrow is usually rich in fat. In one study by Ginzel, et al. reported that 70% of patients with axial tumors due to EMH had evidence of fat components in the on imaging of the masses [5]. Our patient had a diminutive and calcified spleen on CT scan with diffuse osseous sclerosis and multiple biconcave vertebral bodies compatible with a history of chronic anemia and multiple splenic infarcts. A follow-up chest CT scan with IV contrast done five years after the initial diagnosis showed no significant changes in the bilateral soft tissues masses further supporting the diagnosis of EMH.
Discussion

EMH is a compensatory mechanism that occurs in the setting of a hematological disease. Myeloid metaplasia most commonly occurs in the spleen, lymph nodes and liver but it can rarely be seen in other places [6]. Although seldom reported in sickle cell disease and even other hematological abnormalities, PSM due to EMH might be underreported and might be more prevalent than previously thought [7]. Studies of the incidence and prevalence of these soft tissue masses in different hematological diseases are rare and while some reports describe PSM in thalassemia intermedia, data pertinent to these findings in the setting of sickle disease are lacking. In one comparative study by Foroughi, et al. involving 86 patients with thalassemia major and intermedia, 15 patients with thalassemia intermedia had PSM on radiological imaging while none of the β thalassemia major had paravertebral masses [7,8]. All β thalassemia major patients were transfusion dependent compared to the thalassemia intermedia patients, the regular blood transfusions and not hydroxyurea are thought to have prevented the occurrence of PSM [7]. Furthermore there was no difference between the thalassemia intermedia patients treated with hydroxyurea and the group of patients without treatment (7 vs 8; p = 1), hence challenging previously published cases weakly hinting on the efficacy of hydroxyurea in the treatment of EMH [9,10]. The need for treating PSM is questionable. These tumors rarely cause symptoms and the complications attributed solely to presence of PSM are rarely seen. In a study by Dore F, et al. 29 patients with thalassemia intermedia and EMH, 15% had symptoms related to the occurrence of myeloid metaplasia and most resolved upon treating the underlying hematological disease and not by targeting the masses specifically [11]. PSM due to EMH can rarely compress the adjacent vertebral structure causing nerve root compression and paraplegia. Successful treatment with concomitant surgery and radiotherapy has been described. Upon reviewing the literature we have found only one case of paraplegia due to PSM treated solely with radiotherapy, the rest were all surgically treated with or without radiotherapy [12-14].

The absence of histological confirmation may render the diagnosis questionable. However the diagnosis can be safely established when certain radiological features are present in a setting of chronic anemia [15]. Furthermore, these tumors are highly vasculatized and the risk of bleeding from biopsy is significant [16]. Therefore, biopsy should be reserved to cases with signs favoring different etiologies such as the absence of chronic anemia, the presence of bony erosions, the presence of lymphadenopathy, and in symptomatic masses.

Conclusion

Although rarely described, PSM due to myeloid metaplasia in the setting of sickle cell disease can occur. Awareness of this entity is crucial to avoid misdiagnosing these entities. Clinical correlation with a history of chronic hematological disease along with typical imaging features should be helpful in confirming the diagnosis. In our case, the history and radiological findings allowed us to spare the patient an invasive diagnostic test such as biopsy with potential complications such as bleeding. Further studies are needed to elucidate potential therapies and the incidence of these soft tissue masses due to myeloid metaplasia.

References