

An Unusual Cause of the Transient Osteoporosis of the Femoral Head: Essential Thrombocytosis

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Learning Point of the Article:

This case report would like to emphasise the importance of keeping in mind a rare diagnosis, essential thrombocytosis, in femoral head transient osteoporosis.

Abstract

Introduction: Transient osteoporosis (TO), also known as bone marrow edema syndrome, is characterized by marrow edema which causes increasing of intramedullary pressure. Among the other causes, such as 3rd-trimester pregnancy, impaired circulation may also be caused by changes in normal blood counts such as thrombocytosis. There are multiple reports in the literature of secondary femoral head TO transient osteoporosis; however, there is no case report of femoral head TO transient osteoporosis specifically secondary to essential thrombocytosis (ET) in literature. Herein, we present ET essential thrombocytosis as an unusual cause of TO transient osteoporosis of the femoral head.

Case Report: A 33-year-old male presented himself to our clinic with severe left hip pain. The patient has painful hip motions and itching after the bath and episodic chest pain. Family history is positive for hematological malignancy. Complete blood count (CBC) test showed an increased level of thrombocytes and haematology opinion obtained. The genetic analysis was positive for Janus kinase 2 JAK2V617F and patient was diagnosed as ET essential thrombocytosis. Besides, to eliminate the symptoms of the hip, the surgical intervention had planned and the patient had a dramatic pain relief during follow-up.

Conclusion: ET Essential thrombocytosis a myeloproliferative disorder and has a good prognosis. TO Transient osteoporosis could be accompanied with hematologic disorders, especially myeloproliferative diseases. Based on our review of literature, this is the first case with ET essential thrombocytosis and TO transient osteoporosis of the hip.

Keywords: Decompression, essential thrombocytosis, transient osteoporosis. transient osteoporosis, decompression, essential thrombocytosis

Introduction

Chronic myeloproliferative diseases are hematological malignancies characterized by uncontrolled proliferation of myeloid precursor cells. These are, namely, chronic myeloid leukemia, polycythemia vera, chronic myelofibrosis, chronic eosinophilic leukemia, chronic neutrophilic leukemia, mastocytosis, and essential thrombocytosis (ET). About 80% of people with chronic myeloproliferative diseases have a contributing acquired gene mutation. This gene may involve the Janus kinase 2 (JAK2), calreticulin, or MPL gene mutation.

The most common cause of death is due to thrombotic complications [1]. ET is a breakpoint cluster region-abelson(-) chronic myeloproliferative disease that presents with isolated thrombocytosis. It has the longest average survival among chronic myeloproliferative disorders. Patients are usually asymptomatic at the time of diagnosis. In cases of incidentally detected thrombocytosis, causes of reactive thrombocytosis must be excluded. Many conditions, physiological and pathological (infections, neoplasia, tissue damage, etc.) may cause thrombocytosis. Femoral head transient osteoporosis (TO) is a disease that usually affects

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Figure 1: Pre-operative X-rays. (a): Left hip AP. Note that left hip shows osteoporotic changes. (b): Left hip lateral X-ray.

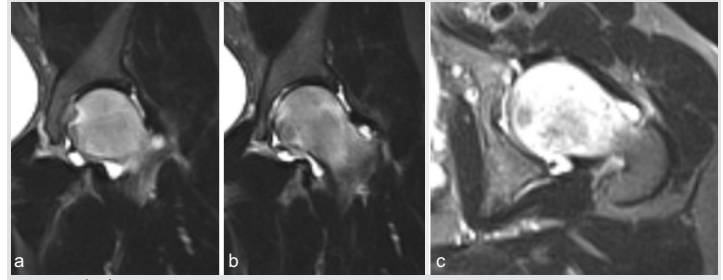


Figure 2: (a-c) Pre-operative left hip non-contrast magnetic resonance imaging shows the high intensity of the femoral head in the T2-weighted image.

people in the third decade. The exact prevalence of TO of the femoral head is unknown. A variety of traumatic and atraumatic factors contribute to the etiology of TO; however, third-trimester pregnancy is associated with most of the atraumatic cases. Avascular necrosis of the femoral head, septic arthritis, and some inflammatory joint diseases should be considered in the differential diagnosis. Mainly it is challenging to distinguish between avascular necrosis of the femoral head and TO of the hip [2]. Some authors declared that the TO of the hip might be an earlier phase of the avascular necrosis of the femoral head [3]. However, it is reported by Yamamoto et al., hematopoietic and fatty elements are affected through ischemic insult in TO of the hip, whereas cell deaths include osteocytes in femoral head avascular necrosis [4]. There are multiple reports in literature of transient secondary osteoporosis of the hip due to chronic myeloproliferative diseases; however, we have not encountered any case report of TO of the femoral head specifically secondary to ET in literature yet. We would like to present ET as an unusual cause of TO of the femoral head.

Case Report

A 33-year-old male patient presented to orthopedics and traumatology outpatient clinic with left hip pain. The pain started suddenly 3 months ago while walking and the patient was conservatively treated. Due to pain did not subside despite conservative therapy, magnetic resonance imaging (MRI) scan was performed. It showed bone marrow edema of the left femoral head, and consequently, the patient was referred to our tertiary clinic to have surgery. Patient's history was significant for palpitations, itching after the bath and episodic chest pain starting approximately 3 months ago. The patient also complained of paresthesias at the hands, feet, and back of the

neck 6–7 months ago and his medical history was unremarkable including steroids, and he did not have any concomitant diseases. Patient's medical and surgical history were unremarkable; however, family history was positive for a hematological malignancy in his sibling. The patient was using crutches. Inspection of the hips and lower extremities was normal. Passive flexion and extension of the left hip elicited pain with normal range of motion. Internal rotation of the left hip was painful and restricted, while external rotation was normal with the full range of motion. The neurovascular examination was unremarkable. The anterior-posterior and lateral X-ray of the hips were taken (Fig. 1). MRI shows increased intensity on T2-weighted images (Fig. 2). Patient's complete blood count (CBC) done at another hospital showed thrombocyte count of 609 103/ μ L whereas other parameters were in normal range. At the time of presentation, CBC was normal except the thrombocyte level (644 103/ μ L). Considering the presenting symptoms and diagnostic tests of the patient, the initial diagnosis was established as ET, hematology opinion obtained. A genetic analysis was positive for JAK2 V617F. Repeated hemograms during the follow-up showed increasing thrombocyte counts (665 103/ μ L and 769 103/ μ L, respectively) which support the diagnosis. To prevent possible thrombotic complications, the patient was started on acetylsalicylic acid and hydroxyurea and the patient has scheduled a bone marrow biopsy. While protein electrophoresis was normal, bone scintigraphy showed significantly increased activity in the left femoral head and acetabular region. The patient was diagnosed with TO of the hip and core decompression was planned to eliminate compressive symptoms of the hip.

Procedure

The surgery was performed on the traction table. An 8 mm drill was used for decompression targeting femoral head (Fig. 3). During the procedure bone marrow, and femoral head biopsies were performed, and the materials were sent for histopathological examination. The results of the per-operative samples came out as "marrow parenchyma including fields of

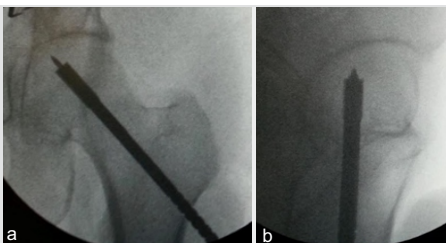


Figure 3: (a and b) Intraoperative core decompression procedure under fluoroscopy.

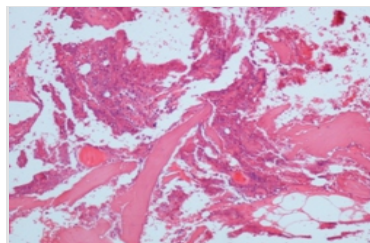


Figure 4: Hemorrhagic necrosis between bone trabeculae.



Figure 5: Post-operative hip X-ray at post-operative 2 years.

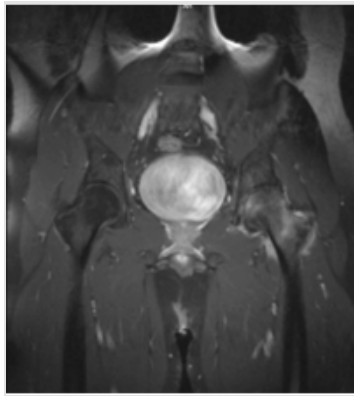


Figure 6: Post-operative hip magnetic resonance imaging

necrosis” and “hemorrhagic necrosis between bone trabeculae” (Fig. 4). The results of the bone marrow biopsy from the posterior iliac crest were “focal megakaryocytic dysplasia.” The symptoms, blood tests, and bone marrow biopsy were consistent with ET which was diagnosed by the hematologist. In the post-operative follow-up of the patient, there was no hip pain at 2 years, and thrombocyte count was 463 103/ μ L. X-ray has no specific changes (Fig. 5), and MRI shows a decreasing of the intensity of edema (Fig. 6).

Discussion

Etis one of the chronic myeloproliferative diseases. The disease usually courses asymptomatic and must be considered in cases of isolated thrombocytosis noticed through routine blood work-up. Before making the diagnosis, the causes of reactive thrombocytosis must be excluded as well. The criteria for ET were revised in 2015 [5] while ET is known as the subtype among chronic myeloproliferative diseases with the best prognosis, thrombotic complications may be, and most of the mortality develops in this background [1]. Signs and symptoms depend on where the clot forms. Our patient has an unusual presentation with severe left hip pain. Blood tests showed high thrombocytes. In the patient’s history, itching after the shower was a clue to consider the ET. A sign and symptoms put together, we considered ET in the orthopedic outpatient clinic initially. The diagnosis was confirmed by the hematologist due to the fulfillment of the WHO criteria [5]. MRI without contrast is the gold standard in diagnosing osteonecrosis of the femoral head [6]. Technetium 99 bone scintigraphy and single-photon emission computed tomography (SPECT) is also useful

in the diagnosis of avascular necrosis; with scintigraphy having a sensitivity of 78–83% and specificity 75–83%, and SPECT having a sensitivity of 92% and specificity of 78%, respectively [7,8]. TO is an uncommon condition among the population. The exact prevalence of the TO is unknown; however, diagnosed more frequently in healthy middle-aged men with a male:female ratio of 3:1 [9,10]. TO is a self-limiting condition, and the treatment in patients with TO of the hip mostly includes conservative modalities. Among non-operative strategies are decreasing weight load comes initially, then bisphosphonates (to decrease osteoclast activity), hyperbaric oxygen, and extracorporeal shockwave therapy treatment. As antilipolytic and anti-coagulant medications statins, anticoagulants and vasodilators can be used. Operative treatment options include core decompression, which the local pressure of the femoral head to extinguish the symptoms. The most commonly employed surgical treatment is core decompression for TO of the femoral head. The theoretical advantage of core decompression is relieving the intraosseous pressure, allowing increased vascularity and reduces edema. While there have been reports TO due to chronic myeloproliferative diseases in literature, we were unable to find any reports of femoral head TO due to ET. Therefore, although rare, ET must be considered in the differential diagnosis of the TO of the femoral head.

Conclusion

To of the hip is a rare diagnosis in middle-aged population and places a significant burden on the workforce. In patients with femoral head TO, underlying conditions must be inquired, and the treatment should take a multidisciplinary approach according to the etiology. ET, one of the rare reasons for the TO of femoral head, should be considered in the differential diagnosis.

Clinical Message

To of the femoral head is seen rarely and often overlooked. In some hematologic disorders; involvement of the femoral head is common and could be the major cause of the disability and patient dissatisfaction. TO of the femoral head should always be kept in mind in patients with hematologic disorders, especially in myeloproliferative disorders.

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