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Case report

CASE REPORT

A case report of a rare omental extramedullary hematopoiesis in an adult: 'an idiopathic finding'

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Abstract

This is a case of a 62-year-old female, known to have multiple medical problems, who presented to her primary care physician with an intermittent abdominal pain and discomfort for a few months. The initial work-up showed mild leucocytosis and a small mass in the omentum. Given that the most concerning differential diagnosis was malignancy, the patient was referred to oncology, where biopsy of the mass showed omentum extramedullary hematopoiesis. The differential diagnosis was wide; however, a repeat computed tomography (CT) scan of the abdomen and pelvis did show persistence of the omental mass. After ruling out any possible causes, including myelofibrosis, with a normal bone marrow, her extramedullary hematopoiesis was deemed of unknown origin and with no clear explanation. Therefore, the patient was diagnosed with a rare adult idiopathic omental extramedullary hematopoiesis that was stable over time.

INTRODUCTION

Hematopoiesis normally occurs in the bone marrow of the normal individual. Extramedullary hematopoiesis (EMH) is a finding that can be found in children and rarely in adults [1]. EMH in adults is mostly an abnormal finding where it is secondary to a certain disease or condition, namely, myelofibrosis or in rare instances chronic hemolytic anemia, infection or inflammatory process or bone marrow irradiation [2].

Usually if EMH were to occur in an adult, its location was well described in the literature and it is most commonly found in the liver, spleen, lymph nodes, kidney, and parasacral areas [3]. However, rare locations for EMH, such as the central nervous system, skin, orbits and omentum, have been described in the literature [2–4].

Idiopathic EMH in the omentum of an adult patient is very rare.

CASE REPORT

A 62-year-old African American female presented to her primary care physician with intermittent abdominal pain and discomfort for a few months. On review of systems, she denied any recent illnesses, nausea, vomiting, diarrhea, fever, chills or weight loss. Besides very faint bilateral wheezing on lung auscultation, her physical examination was quite unremarkable. Her past medical and surgical history consists of asthma, type 2 diabetes mellitus, dyslipidemia, hypertension, previous tobacco user, cholecystectomy in 1988 and tubal ligation in 1986. The patient's family history was negative for any malignancies except for her mother passing away from endometrial cancer and one brother who died from prostate cancer. An initial computed tomography (CT) scan of the abdomen and pelvis showed a 2.1-cm soft tissue mass within the omentum (Fig. 1). There was no hepatomegaly or splenomegaly, no lymphadenopathy or any abdominal

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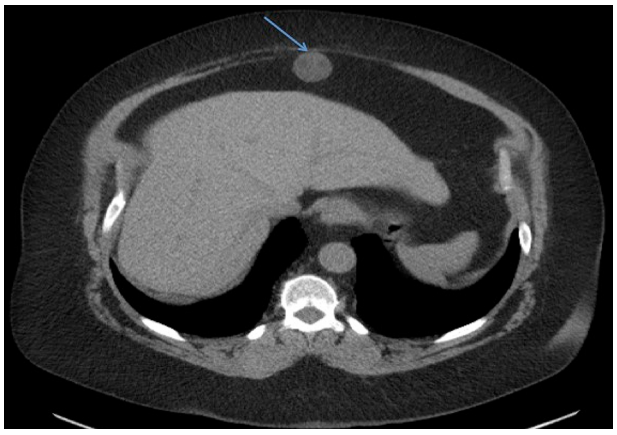

Figure 1: CT scan of the abdomen in axial view, showing 2.1 cm in greatest dimension, within the omentum, just posterior to the anterior wall.

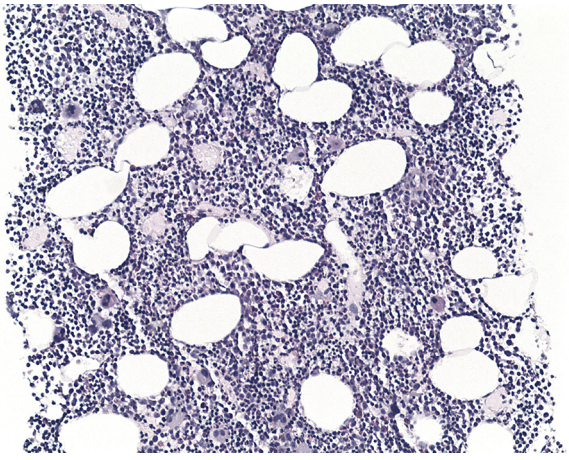

Figure 2: Hematoxylin–eosin stained section (100×): low-power view of omental biopsy, showing fatty tissue with extramedullary hematopoiesis.

effusions described. The differential diagnosis was quite wide and included mainly malignant mass, especially after a repeat CT scan of the abdomen and pelvis 6 months after the initial presentation that showed the mild growth to a 2.4-cm soft tissue mass within the omentum. Therefore, a CT-guided biopsy of the omental mass was persued and showed mature adipose tissue and omentum EMH (Figs 2 and 3). Per all the imaging done and biopsy of the omental mass, no concern or possibility of an accessory spleen was described. At that point, a secondary cause for her EMH was considered with a wide differential diagnosis including myelofibrosis, chronic hemolytic anemia or hemoglobinopathies, inflammation/infection and bone marrow irradiation, which were all excluded with the following investigations: a peripheral smear that did not show any hemolysis and only showed hypochromic red blood cells with mild leukocytosis; routine biochemical tests, including liver, pancreatic and renal function tests, serum immunoglobulins, blood and urinary protein electrophoresis and other tumoral markers were within normal ranges. Then, a bone marrow biopsy and aspirate, with flow cytometry, immunophenotyping and FISH test showed a normal bone marrow (Figs 4 and 5), and BCR/ABL testing in addition to JAK2 on peripheral blood were negative.

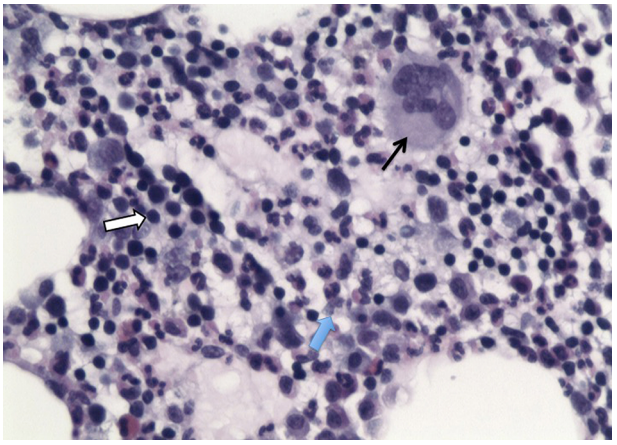

Figure 3: Hematoxylin–eosin-stained section (400×): high-power view of hematopoietic cells (black arrow showing a megakaryocyte, white arrow pointing to erythropoietic cells, and blue arrow depicting myeloid precursors).

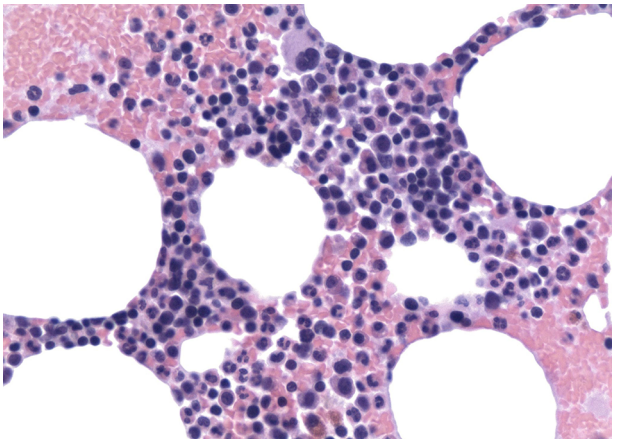

Figure 4: Hematoxylin–eosin-stained section (400×): high-power view of bone marrow showing erythroid and myeloid precursors with megakaryocytes.

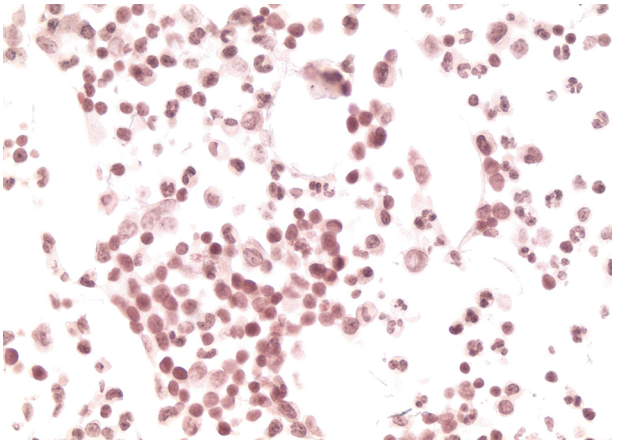

Figure 5: Hematoxylin–eosin-stained section (400×): high-power view of bone marrow particles with reticulin stain showing no evidence of myelofibrosis.

The patient was reassured that her EMH was not secondary to any identifiable cause, and it was deemed idiopathic in nature.

Since the initial presentation, no treatment was given for the patient except supportive care and pain management.

The patient is alive and doing well. Her symptoms have been almost resolved. On serial follow-up for almost 10 months since the first visit, she has been doing better with no significant complaints. Blood counts to date remain stable.

DISCUSSION

EMH's common locations are found mainly in the reticuloendothelial system, and therefore, the most prevalent organs where it can present are the liver, spleen, kidneys, lymph nodes and the parasacral areas [5].

Rare peculiar locations for EMH have also been well described in the literature, and EMH was described in almost any organ and system in the human body [5].

EMH, if found in adults, is almost always secondary to another disorder. Idiopathic EMH with no clear cause has been rarely described [1].

To the best of our knowledge, this is the third case in the literature where EMH was described in the omentum, and only the first one to describe an omental EMH with no clear pathological cause after ruling out all the common disorders and clinical conditions that might explain it [4–6].

In a recent Mayo clinic research paper, Fan *et al*. presented by far the largest experience of EMH without myeloproliferative disorders. They found over 300 cases where almost all causes of EMH were either secondary to hematological or solid malignancies or hemoglobinopathies. Fan *et al*. however identified 12 completely idiopathic EMH where their locations where mostly in the presacral and paraspinal areas or spleen; none were found in the omentum. None of the patients with idiopathic EMH harbored occult malignancies or subsequently developed myeloproliferative disorders or other malignancies. Therefore, their conclusions do not support undertaking extensive investigations targeting myeloproliferative disorders or other malignancies in idiopathic EMH, and simple monitoring might be sufficient [7].

In a previously published report of 27 Mayo Clinic cases of non-hepatosplenic EMH-diagnosed antemortem between 1975 and 2002, the most common associated condition was myelofibrosis, and the most frequent involved site was the thoracic vertebral column [8].

EMH was also associated with solid malignancies, and the diagnosis was made mostly by pathology as part of the diagnosis and staging of the original tumor. The recommended treatment is the treatment of the primary malignancy, and the mortalities were due to the malignant solid tumor rather than EMH [9].

In conclusion, EMH is well described in adults, though it is still a very rare disease.

EMH occurs commonly in the spleen, liver or rarely in the kidneys and the vertebral area and, if found in adults, it is almost always secondary to a primary illness, mainly myelofibrosis. In our unique case, the location of the patient's EMH appeared as a mass in the omentum, a finding that is extremely rare and with no evident cause to explain it that rendered it idiopathic. Idiopathic EMH per the literature will most likely not progress into any form of malignancy, and rather conservative management is best especially for asymptomatic individuals.

The patient in our case did not show any signs of malignancy, and after 10 months did not progress into any other forms of disease, with close follow-up, she will continue to live a normal healthy life with a rare unique finding that so far only needs close monitoring.

ACKNOWLEDGEMENTS

None.

CONFLICT OF INTEREST STATEMENT

None declared.

FUNDING

No financial disclosure to declare.

ETHICAL APPROVAL

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

CONSENT

Written consent has been obtained from the patient.

GUARANTOR

Tarek Haykal, MD, is the named guarantor of the manuscript.

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