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A rare case of extramedullary hematopoiesis in adrenal mass

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ABSTRACT

Extramedullary hematopoiesis (EMH) is a compensatory mechanism for deficient formation or function of red blood cells. Very rarely, extramedullary hematopoiesis is finding out in the kidneys as well as adrenals. We present a 50 year old man, who was admitted in the Urology Department with symptoms of renal colic. A contrast enhanced computed tomography (CT) of the abdomen and pelvis revealed right adrenal lesion of 80/79 mm in coronary size. The final diagnosis was myelolipoma of the right adrenal gland. We presented a rare case of adrenal tumor mass in patient without any haemotological disease that requires surgical excision.

Introduction

Extramedullary hematopoiesis (EMH) is a compensatory mechanism for deficient formation or function of red blood cells. It occurs most often due to hemolytic anemias such as thalasemia, hereditary spherocytosis and sickle cell anemia. It also could be seen in prolonged iron deficiency anemia, myelofibrosis, polycythemia, leukemia and lymphoma. Most commonly it occurs in the spleen, liver and lymph nodes and less frequently it is detected in the lung, pleura, breast, thymus, small bowel and central nervous system. Very rarely, extramedullary hematopoiesis is finding out in the kidneys as well as adrenals.

Case presentation

We present a 50 year old man, who was admitted in the Urology Department with symptoms of renal colic. Laboratory investigations showed hemoglobin 122 G/L, WBC count 7.4×10^9 G/L, platelet count 180×10^9 G/L, MCV 64.7 fl, MCH 20.8 pg. The results from biochemical investigations were: serum bilirubin 13.5 μmol/l, conjugated bilirubin 2.5 μmol/l, blood urea 5.5 mmol/l, serum creatinine 112.0 μmol/l, serum calcium 2.0 mmol/l, serum potassium 5.6 mmol/l, serum sodium 135.0 mmol/l, alanine aminotransferase 26.1 U/I, aspartate aminotransferase 21.3 U/I. Tests for hepatitis C virus (anti HCV antibody) and HIV antibodies (Anti HIV-1,2), serum HBsAg and HBc antibody were negative. Urinalysis in sediment, showed many leukocytes and erythrocytes.

From abdominal sonography left suprarenal solid mass in size of 8.5 × 8.2 cm, with heterogenous ecostructure was detected. A contrast enhanced computed tomography (CT) of the abdomen and pelvis revealed right adrenal lesion of 80/79 mm in coronary size- heterogeneous with presence of adipose tissue centrally up to 60 HU and denser sections peripherally up to 40 HU, replacing the gland almost completely (Fig. 1).

Because of nephrosclerosis of the right kidney and loss of function, a laparoscopic right radical nephron-adrenalectomy was performed. Macroscopically encapsulated tumor with a soft consistency of 9 cm in diameter was found.

Histopathologic evaluation revealed a tumor mass composed of mature adipose tissue with extensive hemorrhages, including haemopoietic islands with megakaryocytes. Lymphocytic infiltrates are established to the extent of lymphatic follicles with germinal centers and foci of necrosis. The final diagnosis was myelolipoma of the right adrenal gland (Fig. 2).

Control examination and CT scan was recommended 6 months after surgery and also monthly evaluation of his peripheral blood count.

Discussion

EMH is usually asymptomatic and discovered incidentally. Symptomatic cases occurs due to mass effect with compression to adjacent organ. The surgical indication for excision of the adrenal incidentalomas is the tumor size. Adrenal tumor larger than 6 cm in diameter must be excised. In these cases, the risk of adrenal cancer is 35%–98%.[1,2] In our case the size of the tumor was 9 cm in diameter.

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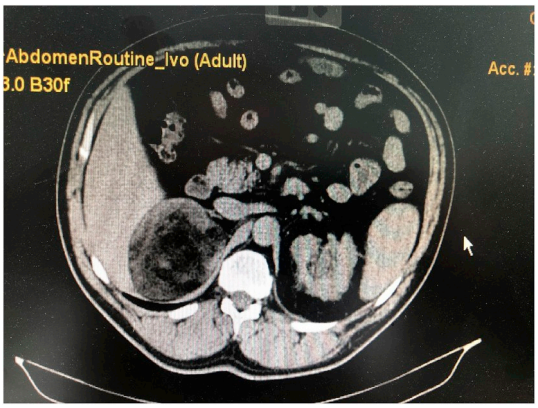

Fig. 1. CT scan of tumor mass at the right adrenal gland.

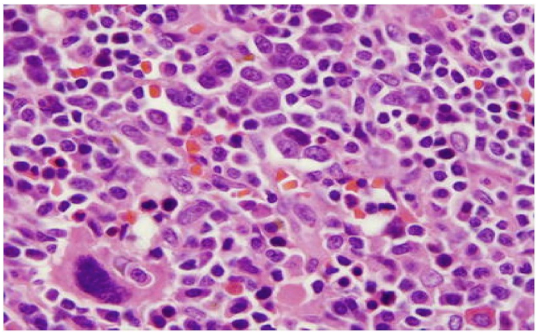

Fig. 2. Histopathology image of EMH in adrenal.

The adrenal glands are the seat of a wide variety of diseases. However, EMH in the adrenal is uncommon and it is thought to be a compensatory, physiological mechanism that occurs during altered medullary haematopoesis which is commonly seen in the haemoglobinopathies, leukemias, lymphomas and myelofibrosis.[3,4] In our case the patient do not have any haemoglobinopathies.

The exact mechanism of EMH in the adrenal gland is unknown, but several hypotheses are suggested. The adrenal gland has hematopoietic capacity during the fetal period and EMH may develop from primitive rests in diseased condition. Other scientists believe that embolization of hematopoietic stem cells and homing in adrenal gland may occur. Chronic hypoxia is another presumptive cause of EMH.[5] Our patient has only a positive history of anemia in the past.

Conclusions

We presented a rare case of adrenal tumor mass in patient without any haemotological disease, that requires surgical excision. In the setting of a hematological disorder, a biopsy is indicated in the management of an adrenal incidentaloma. If it reveals extramedullary haematopoesis, excision can be avoided.

Declaration of competing interest

The authors declare that they have no competing interests.

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