Adrenal extramedullary hematopoiesis: Report on a pediatric case and update of the literature

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Abstract. The authors report on a rare pediatric case of adrenal extramedullary hematopoiesis in a patient with beta-thalassemia disease. The lesion was clinically discovered as incidentaloma of the right adrenal gland and treated by surgery. Adrenal extramedullary hematopoiesis may clinically be detected as incidentaloma. Adrenal incidentalomas presenting with hematologic disorders, such as agnogenic myeloid aplasia and beta-thalassemia, need careful imaging as well as adrenal hormonal investigation in order to exclude malignancy and subclinical hypersecretory syndromes. Ultrasound or CT-FNA of the lesion are effective in finding out the disease.

Key words: Adrenal, Extramedullary hematopoiesis, Incidentaloma, Thalassemia

Introduction

Extramedullary hematopoiesis arises in organs which are provided of pluripotential stem cells. Most commonly it occurs in the spleen, liver and lymph nodes; less frequently it is detected in the lung, pleura, breast, thymus, small bowell and central nervous system. Very rarely, extramedullary hematopoiesis is find out in the kidneys [1–4] as well as adrenals [5–10]. Herein we report on a rare pediatric case of adrenal extramedullary hematopoiesis which was clinically detected as incidentaloma.

Case report

A 10-year-old caucasian female with beta-thalassemia was admitted with a right adrenal mass which was incidentally detected elsewhere by sonography. Physical examination revealed splenomegaly. Hemoglobin was 10 g/dl. The read blood morphology and the electrophoretic pattern of hemoglobin were typical of beta-thalassemia disease. Sonography showed the right adrenal gland with a well-defined, hypoechoic, round mass which appeared non-homogenous, hypodense, and sized 4.5 cm on computed tomography (CT). Adrenal hormonal investigations resulted normal. It was concluded as follows: nonhypersecretory right adrenal incidentaloma in patient with beta-thalassemia disease. The patient underwent right adrenalectomy by the subcostal transperitoneal approach. The postoperative period was uneventful. Gross examination of the specimen showed the adrenal gland including a brownish mass which was sized 5 cm in diameter. Histologic examination of the sections demonstrated normal adrenal parenchima surrounding extramedullary hematopoiesis (Figure 1a) which contained hematopoietic cells at various stages of maturation with predominance of the erythroid series (Figure 1b). The patient died 72 months later because of disease’s complications related to infection and heart failure.

Discussion

After a research of the English literature concerning this subject, we find out two reports of adrenal extramedullary hematopoiesis [8, 9]. Adrenal ectopic haemopoiesis is associated with hematologic disorders including agnogenic myeloid metaplasia, beta-thalassemia and myelofibrosis [5–9]. After including this report, the female/male ratio is 2:1 and the average age is 35 years (range 10–66). Herein we report the
Figure 1. Adrenal extramedullary hematopoiesis. Normal adrenal parenchima surrounding extramedullary hematopoiesis (a) including hematopoietic cells at various stages of maturation with predominance of the erythroid series (b).