Turk J Surg 2017; 33: 116-118
DOI: 10.5152/UCD.2015.2983

Case Report

Extra-adrenal myelolipoma with hemolytic anemia

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ABSTRACT

Myelolipomas are rare benign tumors often detected as adrenal masses. Extra-adrenal myelolipomas are encountered even more rarely. The rate of detection of these lesions is increasing with improved radiological techniques. Because of their localization and morphological similarities to well differentiated liposarcomas, extra-adrenal myelolipomas need to be differentiated from other aggressive neoplasms. Preoperative imaging and percutaneous biopsy are important tools in the diagnosis of these lesions. We report a very rare case of an extra-adrenal perirenal myelolipoma associated with hemolytic anemia. The etiology, differential diagnosis, and treatment options for the lesion have been discussed. Fat-containing tumors of the retroperitoneum should be considered in the differential diagnosis. Accurate diagnosis is important to avoid over-treatment of these benign lesions.

Keywords: Myelolipoma, anemia, hemolytic

INTRODUCTION

Myelolipomas are small, asymptomatic, infrequently encountered lesions of the adrenal cortex (1). They frequently contain mature adipose tissue and hematopoietic elements (both myeloid and erythroid) (2). Extra-adrenal myelolipomas are rarer, with about 100 cases reported in the literature most of which are associated with different lesions and encountered in diverse localizations (2). The incidence of myelolipomas in the autopsy series is less than 1% (3). To the extent of our knowledge, no case of extra-adrenal myelolipoma with accompanying hemolytic anemia has been reported. The known localizations are the presacral soft tissue, retroperitoneum, pelvis, stomach, and rarely the perirenal tissue (4-7). The prognosis is very good and no malignant degeneration has been reported (8). The histological appearance of extra-adrenal myelolipomas is identical to that of well-differentiated liposarcomas. Therefore, retroperitoneal tumors with fat content should always be kept in mind during the differential diagnosis.

CASE PRESENTATION

A 26-year-old woman presented to our hospital with abdominal pain and malaise. There was no significant knowledge of her medical and familial history. Abdominal ultrasonography and abdominal computerized tomography (CT) revealed an 8 cm perirenal mass, not related to the adrenal gland (Figure 1a, b). Her laboratory values showed anemia with iron and vitamin B12 deficiency [Hemoglobin: 6.2 gr/dL, N: 12.2–18.1 gr/dL; mean corpuscular volume: 81.1, N: 80–96; and the red cell distribution width: 14.4, N: 11.8–15.6]. The white blood cell and platelet counts were within normal ranges. Detailed laboratory tests were performed and elevated C-reactive protein and ferritin values, as well as indirect Coombs' test positivity, and normal liver and kidney function tests were determined. To exclude an infectious etiology, blood and urine cultures and tube agglutination tests for brucella and salmonella were performed, all of which turned out to be negative. The tumor markers (CA 125, CA 19-9, CEA) were within the normal range. Following the hospitalization of the patient, the daily complete blood count analyses showed a gradual decrease in the hemoglobin levels to 3.8 g/dL. Empirical antibiotic treatment (ceftriaxone 2 g and metronidazole 1.5 g daily) was initiated. The peripheral blood smear of the patient was compatible with partial hemolysis. Bone marrow aspiration and smear and other laboratory tests were performed and were found to be compatible with hemolytic anemia. Intravenous prednisolone was initiated and blood replacement was performed. Under oral deltacortril treatment (10 mg/day), her hematological parameters had a stable course. We decided to perform laparotomy because of a symptomatic (pain and hemolytic anemia) mass in the abdomen. Before the operation, we placed a double J stent in the right ureter. Conventional surgery was performed. In the right perirenal area, we observed a mass 8 cm in diameter encircling the right ureter. The ureter's wall was observed to be very thin. The mass was excised totally with conservation of the right ureter, kidney, and adrenal glands. After the surgery, the deltacortril dose was gradually reduced and then stopped on the 20th day. The patient was discharged and followed-up in the out-patient clinic and her consent for publication was taken. Her hematological parameters were observed to be stable.

Cite this paper as:
İflazoğlu N, Üreyen O, Keleş M. Extra-adrenal myelolipoma with hemolytic anemia. Turk J Surg 2017; 33(2): 116-118

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Received: 08.11.2014
Accepted: 21.12.2014
Available Online Date: 06.07.2015

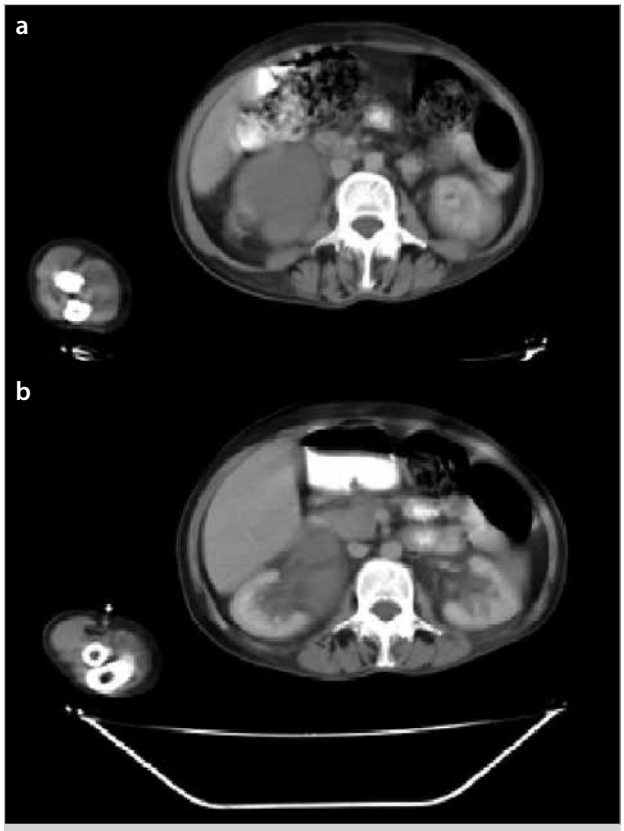

Figure 1. a, b. (a) Perirenal mass, computerized tomography. (b) Perirenal mass, computerized tomography

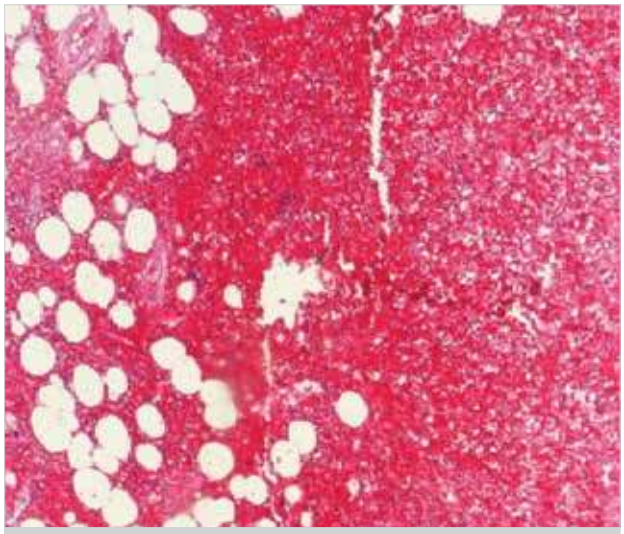
Figure 2. Fat and myeloid cells, hematoxylin–eosin, ×100

The excised mass was evaluated histopathologically. A 8 × 6 × 6 cm mass having an intact capsule and external appearance that was partly yellow and partly brown was observed. Dispersed among the fat cells were non-uniform hematopoietic areas containing granulocytic, erythrocytic, and megakaryocytic elements (Figure 2). In the light of the pathological findings, the mass was diagnosed to be a myelolipoma.

DISCUSSION

Myelolipomas are rare benign tumors of mature hematopoietic tissue and fat and tumorigenic processes are poorly understood. They most commonly occur in the adrenal glands, but extra-adrenal myelolipomas have been reported in other locations such as the presacral region or retroperitoneum (4-7). It is not unusual that they are incidental findings revealed during the evaluation for different diseases (9). Although there are several theories regarding their etiology, the process is not yet fully understood. One of the theories proposing these tumors is the result of the differentiation of ectopic adrenal or hematopoietic cells in response to a triggering stimulus (10), whereas another theory by Chang et al. (11) suggests that myelolipomas are formed as a result of translocation t(3;21)(q25;p11). Bishop et al. (12) demonstrated chromosome X inactivation in the fat and hematopoietic cells and proposed a clonal origin for myelolipomas.

Extra-adrenal myelolipomas are observed frequently over the age of 40 with a female predominance. Their diameters are on average about 8 cm, although their size can vary between 4 cm and 15 cm (13), with cases having been reported of being up to 27 cm in size (10). Despite our patient being younger, her sex and tumor size were in accordance with the literature. Extra-adrenal localization can be in any part of the body, but the presacral area and more rarely the retroperitoneum are the most common sites reported, whereas only a few cases have been reported in the perirenal area (2, 10). They are usually incidental findings because they are mainly asymptomatic; only in 10% of cases, depending on the size and site of the lesion, they are observed to be symptomatic due to the compression of local structures (2). Hemorrhage in extra-adrenal myelolipomas is a very rare complication, and no case has been reported in the literature, which caused the clinical findings of acute hemorrhage. Myelolipomas are hormonally inactive, but about 10% are associated with several endocrine disorders such as Cushing's syndrome, congenital adrenal hyperplasia, Conn's syndrome, pheochromocytoma, hyperparathyroidism, or adrenogenital syndrome (2). Our case was diagnosed as a result of evaluation for abdominal pain and serious anemia, and had no other accompanying disorders, endocrinological or otherwise. Hemolytic anemias caused by thalassemia major, thalassemia intermedia, sickle cell anemia, and hereditary spherocytosis have been reported in cases of large adrenal myelolipomas (14, 15), but no case of hemolytic anemia in extra-adrenal myelolipomas have been defined in the literature.

A common consensus on the surgical approach to myelolipomas has not yet been reached due to the limited number of extra-adrenal myelolipoma cases and the literature has thus been limited to case reports. Commonly, surgical excision is performed because of the symptoms that are caused by mass effect and due to the fact that the nature of the tumor is unknown (2). Adrenal myelolipomas can usually be diagnosed radiologically, but such is not the case with extra-adrenal myelolipomas due to a lower fat content and rarity of the lesion. In such cases, percutaneous fine needle biopsy can be performed in a simple, safe, and efficient manner (2). Because the mass in our case was symptomatic and sufficiently large, the patient was operated rather than biopsied. Microscopically, extramedullary hematopoietic tumors have a predominance of hematopoietic elements, with erythroid hyperplasia. Fat is not an enlarged component of the process (13). Extra-adrenal myelolipomas may have a predominance of either the hematopoietic or fatty component, usually the latter, and generally have a more conspicuous lymphocyte population (10).

Despite the fact that the presence of hematopoietic cells (myeloid, erythroid, and megakaryocytic) and mature adipose tissue of varying amounts are used for the diagnosis, the observation of megacaryocytes forms the basis of the diagnosis of extra-adrenal myelolipoma (2, 13). Our case demonstrated all three types of the hematopoietic cells as explained above.

CONCLUSION

Consequently, today, it is possible to reach a definitive diagnosis using advanced radiological techniques and percutaneous biopsy; therefore, the number of unnecessary laparotomies and the emotional and economic burden on the patient and the society can be reduced. It must be kept in mind that myelolipomas can, though rarely, cause severe hemolytic anemia and can be located in an extra-adrenal site, and in any part of the body.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - N.İ., M.K.; Design - N.İ., O.Ü.; Supervision - N.İ.; Analysis and/or Interpretation - O.Ü., M.K.; Literature Review - N.İ.; Writer - N.İ., M.K.; Critical Review - O.Ü.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

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