Adrenal Myelolipoma: A Case Report with Literature Review

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A 63-year-old man was admitted to the ward of Internal Medicine Department due to an incidentally found right retroperitoneal tumor under abdominal sonography without any specific discomforts. After further laboratory biochemical investigation and abdominal computed tomography, a right suprarenal mass with fat component density and 7.1 x 6.6 cm in size was detected. So he was transferred to Urology Department for tumor excision under the impression of right adrenal tumor. A final diagnosis of adrenal myelolipoma was confirmed by histopathological studies. This unusual case is reported with a review of the literature. (J Urol R.O.C., 11:185-9, 2000)

Key word: myelolipoma.

INTRODUCTION

Adrenal myelolipomas are relatively rare tumors and only 230 cases have been reported up to date. The tumors usually originate from the adrenal cortex and histologically consisting of mature fat and haemopoietic tissue [1]. They account for 7 to 15% of all adrenal “incidentalomas” [2]. Adrenal myelolipomas are non-functioning and benign in origin, but hormonally active tumors or co-existence with malignancy have been reported. Although diagnosis is not difficult, either pre- or post-operatively, the therapeutic strategy is still controversial. Herein we present a case of adrenal myelolipoma and discuss its clinical presentation, evaluation and management.

CASE REPORT

A 63-year-old male patient who suffered from old cerebrovascular attack with left side weakness, hypertension and hyperlipidemia for more than 10 years with regular medication control was admitted to the Internal Medicine Department for further evaluation due to a right abdominal mass as detected by abdominal sonography in routine health examination. Throughout his whole admission course in the Internal Medicine Department, no general malaise, no poor appetite nor body weight loss were observed. Defecation was also normal without abdominal pain. Only frequent urination was noticed due to moderate prostate enlargement.

The abdominal computed tomogram revealed a right suprarenal mass, fat component in density, 7.1 x 6.6 cm in size with well-defined contour and highly suspected of adrenal origin (Fig. 1).

The laboratory biochemical investigation revealed normal results including blood sugar, blood urea nitrogen, serum creatinine, cholesterol, triglycerides, alkaline phosphatase, serum glutamic oxaloacetic transaminase, glutamic pyruvic transaminase and total bilirubin. Normal values of bleeding time, prothrombin time and partial thromboplastin time were also detected. Hematological study was also within normal range. Levels of adrenal hormones were: cortisol, 8.31 μg/dl at 8 AM (normal: 1-39μg/dl ) and 3.57μg/dl at 4 PM (nor-

Fig. 1 Abdominal CT reveals a right suprarenal mass (arrowhead), fat component in density, 7.1x6.6 cm in size with well-defined contour and highly suspected adrenal origin.
mal: 3-18 μg/dl). In 24-hour urine, there appeared vanillylmandelic acid, 3.28 mg (normal: 1.0 - 7.5 mg); norepinephrine, 42.28 μg (normal: 11.1 - 85.5 μg) and epinephrine, 3.07 μg (normal < 22.4 μg). Furthermore, the levels of tumor markers were: carcinoembryonic antigen, 4.01 ng/ml (normal: 0 - 4.6 ng/ml), and alpha-fetoprotein, 0.93 ng/ml (normal: 0 - 15 ng/ml).

After complete study and evaluation, he was transferred to the Urological ward for further surgical management. Transperitoneal excision of the tumor was performed through right subcostal exploration. The tumor was well-encapsulated and closely adhering to the right adrenal gland. It was totally removed with the right adrenal gland, and the abdomen was otherwise free of tumor. The patient remained well three months postoperatively, and the followup computed tomogram revealed no tumor recurrence, but the hypertension still persisted.

Grossly, the specimen submitted consisted of more than ten tissue fragments measuring up to 11.0 × 9.5 × 1.0 cm in size. They were red-yellowish, fragile and glistening tissue. Some normal thin adrenal tissue was also identified. Microscopically, it showed a picture of myelolipoma of the adrenal gland which is largely replaced by mature adipose tissue (Fig. 2A) mixed with unremarkable hematopoietic elements (Fig. 2B). No evidence of malignancy was seen.

DISCUSSION

Adrenal myelolipoma was first reported as “myelo-adipose formation” or “myelolipoma” in 1966 [3]. It is a non-functioning and benign neoplasm originating in the adrenal cortex and histologically consisting of mature fat and haemopoietic tissue [4]. Although some theories about their origin have been put forward, a lot of things were yet still unclear, including development from the rests of mesenchymal stem cells, the extramedullary hematopoiesis, the embolism of bone marrow, and moreover, according to the most widely accepted therapy, the metaplasia of the reticuloendothelial cells of blood capillaries [5]. The incidence of adrenal myelolipomas at autopsy has been reported to be 0.2% [3], while due to the widespread application of noninvasive, high resolution imaging techniques, the relative diagnostic frequency of myelolipoma as the adrenal “incidentalomas” is 7% to 15% [2]. The sex distribution is female predominant, and the patients range from 30 to 76 years of age [6]. Most tumors originated from the right adrenal [6], and bilateral synchronous tumors have ever been cited as well [5-7]. The patients are usually asymptomatic. In symptomatic patients, the most common symptoms observed are nonspecific abdominal pain, hematuria and hypertension [8]. Some isolated cases of hypersecreting function have been reported even though most of them are hormonally inactive [9-11].

Interestingly, although myelolipomas have been commonly found in adrenal glands, rare extraadrenal myelolipomas (EMLs) are well documented. They have been found in various sites, including mediastinum, liver, stomach, lungs, pelvis, spleen, retroperitoneum, presacral region, thoracic spine and mesentery. EMLs must be told from extramedullary hematopoieses, which are also composed of hematopoietic elements but may lack adipose tissue, and are associated with anemia and marked bone marrow hyperplasia [12].

Imaging studies such as computerized tomographic (CT) scan and magnetic resonance imaging (MRI) are relatively specific and has made the differentiation of adrenal myelolipomas from other retroperitoneal tumors easy. Adrenal myelolipomas typically manifest as a well-defined suprarenal mass with an attenuation of –30 to –115 HU on CT scan [13], and the low density, macroscopic lipid content is characteristic. On MRI, the fatty component appears hyperintense on T1-weighted and intermediate in signal on T2-weighted sequences [14]. These diagnoses shall be made with caution, especially when the lipid content is not predominant due to the possible association with an adenoma.

**Fig. 2A** Myelolipoma of the adrenal gland which is largely replaced by mature adipose tissue mixed with hematopoietic elements, adrenal tissue with atrophic change is well identified (arrowheads). (H & E reduced from ×50)

**Fig. 2B** Hematopoietic elements within myelolipoma composing of hematogeneous cells (arrowhead) and megakaryocyte (arrow) are identified. (H & E reduced from ×250)
Fine needle biopsies under ultrasonic, fluoroscopic or CT guided aspiration have been reported [12,13,15], and they were proved to be useful to establish the definitive diagnosis and select optimal treatment.

Pathologically, adrenal myelolipoma is a nonfunctional benign tumor constituted by adipose tissue and small islands of hematopoietic elements, including the myelotic, lymphocytic, erythrocytic, and megakaryocytic cells [7]. Four clinicopathologic patterns according to CT findings were described by Kenney et al. [16] including (a) isolated adrenal myelolipoma without other disorders, (b) myelolipoma with hemorrhage, more common in larger lesion (diameter > 10 cm), (c) extra-adrenal myelolipomas being similar to adrenal myelolipomas, found most often in the retroperitoneum, and (d) myelolipomatous foci within other adrenal pathologic conditions which present smaller, lower fat content, and are more heavily calcified.

Therapeutically, the strategy is still controversial. Most authors suggested that myelolipoma, if correctly diagnosed, can be treated conservatively with careful followup, while their surgical management is dependent on size or symptoms [7,17]. Just like the therapeutic implications of the adrenal incidentalomas, hormonally active myelolipoma and those suspected for combining with malignancy should be treated surgically, and masses greater than 6 cm should also be removed [15]. However, some authors still suggested that surgical exploration remains a compulsory step in myelolipoma management and its simple removal seems to be the therapy of choice [18].

Open adrenalectomy has been the standard procedure for the surgical removal of adrenal myelolipoma, while some authors have suggested that laparoscopic adrenalectomy can be equivalent to open adrenalectomy in efficiency with a shorter convalescence [19,20].

Although the potential for recurrence or malignant transformation of this benign tumor is rare, post-operative followup with cross-sectional images is still considered necessary. In this case, the patient presented initially without specific discomforts, his adrenal tumor was found incidentally, and the final diagnosis was proved pathologically.

In conclusion, adrenal myelolipomas are rare and always benign adrenal tumors. Radiological imagings, such as CT and MRI, are usually used to characterize a lesion, and are specific enough to provide a preoperative diagnosis of adrenal myelolipoma. Similarly, a needle biopsy is reliable. Open or laparoscopic adrenalectomy is efficient for managing the tumors. Its prognosis is excellent with rare recurrent potential or malignant transformation. Although rare, adrenal myelolipoma should be considered to be a critical stage for differential diagnosis of adrenal tumors.

REFERENCES
