ADRENAL MYELOLIPOMA IN A HEMODIALYSIS PATIENT – CASE REPORT AND REVIEW OF LITERATURE

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Myelolipoma is an uncommon adrenal tumor composed of a variable mixture of mature adipose tissue and hematopoietic elements that resemble the content of bone marrow. Adrenal glands are the commonest involved site, but other extra-adrenal sites are also reported. Myelolipoma is a benign lesion, usually small and unilateral, asymptomatic, and discovered by accident or at autopsy. Most myelolipomas do not cause an endocrine disorder, but cases associated with endocrine disorders have been reported. We report a 63-year-old female who had received regular hemodialysis for two years and the dialysis courses were smooth. She received abdominal ultrasonography to evaluate the source of fever and right-upper-quadrate abdominal pain and abnormal liver function. Accidentally, it revealed an 8.7×5.0 cm lesion over right suprarenal area. Abdominal computed tomography showed a fat-density, homogeneous right adrenal mass. No other intra-abdominal lesions, except bilateral contracted kidneys, could be noted. She was finally operated and histological examination showed that it was mainly composed of mature adipocytes. Hematopoietic cells, resembling the picture in bone marrow, were noted between the fat cells. To our knowledge, this is the first case report in uremic patient. Follow-up examinations showed no recurrence and patient stayed well on hemodialysis. (Acta Nephrologica 2001; 15: 118-121)

Key words: myelolipoma, adrenal gland, uremia.

INTRODUCTION

Adrenal myelolipoma is a non-functioning, benign tumor composed of mature fat tissue and hematopoietic elements, which resemble the bone marrow. Gierke first described such a lesion in 1905,1 but the term “myelolipoma” was first used by Oberling in 1929.2 However, these tumors were most frequently discovered at autopsy and the incidence was estimated to be between 0.08% and 0.25%.3,4 With the widespread uses of noninvasive imaging techniques, particularly ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI), the previously incidental detection of myelolipoma is becoming more common. In one Japanese study, 7% in two hundred and ten adrenal tumors were myelolipomas.5

Myelolipomas rarely exceed 5 cm in diameter, and are commonly asymptomatic. However, larger tumors may cause symptoms due to compression to the surrounding organs. Most tumors do not by themselves cause endocrine disorders, but myelolipoma associated with endocrine disorders have been reported, such as Cushing’s syndrome, congenital adrenal hyperplasia, and Conn’s syndrome.6,7 The lack of obvious symptoms explains the reason why it is not reported in uremic patients, as in this article. The clinical, radiological, and pathologic characteristics of this case are described.

CASE REPORT:

A 63-year-old female was admitted to our hospital due to fever and abdominal discomfort over her right upper quadrant area for uncertain duration. She was a uremic patient from chronic glomerulonephritis. She was also a hepatitis C patient. No other systemic disease, such as hypertension or diabetes mellitus, was noted. She had received regular hemodialysis, three times per week, four hours per session, for 2 years at our hospital. Three to four days before admission, fever developed once again with symptoms of upper respiratory tract infection. Ab-
Abdominal ultrasonography was performed to survey abnormal liver function, shown by routine monthly blood test AST 220 IU/L and ALT 289 IU/L, and the concomitant dull discomfort over right upper quadrant area. Both her kidneys were contracted and there was no obvious lesion in the liver, except chronic change due to hepatitis. However, a hyperechoic heterogeneous tumor, 8.5×5.0 cm, in the right supra-renal area was noted (Fig. 1A). Her fever and upper respiratory tract infection subsided after certain management. She was then admitted for detailed survey on this accidentally found adrenal tumor.

After admission, serial examinations were arranged. She was a 157 cm, 55 kg, slightly obese woman with blood pressure 110/70 mmHg, pulse rate 76 beats per minute. Her conjunctiva was mildly anemic. No abnormal lymph node could be palpated in her neck, axillary, or inguinal areas. Chest auscultation was generally normal. Surprisingly, we were not able to palpate any mass lesion in her abdomen, even in the right upper quadrant area. Murphy’s sign was slightly positive. Her hemoglobin was 9.2 g/dl, albumin 3.8 g/dl, total bilirubin 0.9 mg/dl. Blood electrolyte levels were within normal limits. Measurement of hormones level, including aldosterone, cortisol, ACTH, plasma renin activity (PRA), showed normal values. Abdominal CT revealed a fat-density mass over right adrenal area, and the lesion could not be enhanced after contrast injection (Fig. 1B). Under the impression of right myelolipoma with clinical symptom, surgical intervention was suggested.

Surgery was performed through right flank incision, a well-capsulated 8.5×6×2.6 cm mass, weighing 93 grams, was removed. Grossly, it was grayish yellow and greasy. The cutting surface of the tumor showed several punctate hemorrhagic spots. Microscopically, this tumor was almost completely filled with well-differentiated mature adipocytes (Fig. 2A). Hematopoietic elements, including myeloid/erythroid precursors and megakaryocytes, occupied the interstitial spaces. Atrophic change of the adrenal cortex, resulting from tumor compression was noted (Fig. 2B). After operation, the condition was stable and she was discharged one week later. Follow up ultrasound examination did not show recurrent or new lesion, and no symptom related to adrenal insufficiency was noted after operation. Normal cortisol and aldosterone level were detected 6 months later.

**DISCUSSION**

Myelolipomas are uncommon, benign, and usually asymptomatic adrenal lesions, and are usually discovered incidentally. The first symptomatic adrenal myelolipoma was reported in 1975 by Dyckman and Freedman. They are usually unilateral and the size...
Clinical symptoms may manifest if the tumor sizes are large enough to cause compression or displacement effects. They exhibit variable growth rate and tumor size does not necessarily correlate with symptoms. The most commonly complained symptoms, if present, are soreness or pain in the right flank or subcostal areas. However, the most serious manifestation may be spontaneous retroperitoneal hemorrhage that has been reported with the presentation of shock. One study found that the risk of hemorrhage became much higher when the myelolipoma grew to larger than 10 cm in diameter. Although myelolipoma is usually an isolated, unilateral adrenal lesion, it also occurs in extra-adrenal sites, most commonly in the presacral and retroperitoneal areas. Their clinical and imaging characteristics are similar to those of adrenal glands in situ. Myelolipomatous foci can also be found within other pathologic conditions in the adrenal glands, including hyperplasia and adenomas.

Most myelolipomas are hormone-inactive lesions, but they are frequently associated with obesity and hypertension. A total of twenty myelolipoma cases associated with endocrine dysfunctions have been reported in the English literature. Among them, congenital adrenal hyperplasia caused by 21- or 17-hydroxylase deficiency and Cushing’s syndrome are the two most commonly described endocrine disorders. In addition, primary hyperaldosteronism, pheochromocytoma, and adrenal insufficiency, have also been reported.

Myelolipomas consist of mainly mature fat tissue and hematopoietic elements, including erythroid/myeloid precursors, megakaryocytes, and lymphocytes. They are distinct from true bone marrow because neither reticular sinusoids, nor bone spicules, are present. The origin of adrenal myelolipoma remains unclear. Theories have been proposed, including development from embryonic mesenchymal rests in the adrenal glands, development from hematogenously seeded bone marrow emboli, and metaplasia of reticulo-endothelial cells as a result of chronic stress.

In the past, most myelolipomas were discovered at autopsy and the incidence was low, between 0.08% and 0.25%. With the widespread use of noninvasive imaging techniques, particularly ultrasonography (US), computed tomography (CT), and magnetic resonance imaging (MRI), incidental detection of myelolipoma is becoming more commonplace. On ultrasonography, it typically shows a high-echogenic heterogeneous supra-adrenal lesion. Differential diagnosis includes retroperitoneal lipoma or liposarcoma, exophytic renal angiomyolipoma, adrenal adenoma, adrenal metastasis, and primary adrenal malignancy. Histological analysis is necessary, therefore. However, biopsy may be hampered if hematopoietic elements predominate, or hemorrhage exists. Under such circumstances, computed tomography (CT) seems to be a more valuable technique to help making differential diagnosis. By CT, myelolipoma has a characteristic pattern of low density by dense fatty tissue. Another frequently used non-invasive imaging technique is magnetic resonance imaging (MRI). On MRI, myelolipoma is usually revealed as a homogenous mass with high intensity equal to that of retroperitoneal fat on T1-weighted image or a heterogeneous mass with areas of fat intensity and varying amounts of intermediate signals on T2-weighted images. If the CT/MRI diagnosis is equivocal, fine needle aspiration may be used for confirmation, particularly if hematopoietic element can be identified.

Once identified, follow-up for myelolipoma is im-
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Important because surgery may not be necessary for a small (smaller than 4 cm) and asymptomatic tumor. Surgical approach may then be considered if the tumor enlarges or becomes symptomatic. Primary surgical excision is indicated, in the first place, for symptomatic cases or for large tumors (larger than 10 cm), or when its nature is unclear. In cases with undetermined nature, an echo-guided fine needle aspiration cytology examination is needed for diagnosis. Besides, endocrinological evaluation is indicated, especially before operation, to rule out any associated endocrine disorders.

To our knowledge, this is the first report of myelolipoma in an uremia patient. Abdominal ultrasonography rapidly identified this benign tumor with clinical manifestation of right upper quadrant soreness. CT scanning further approved our clinical suspicion of myelolipoma by its rich fat content. She received surgical resection of this tumor because it was symptomatic. Histological analysis proved the diagnosis by showing mature adipose tissue and interspersed hematopoietic precursor cells. No recurrence has been noted in the follow-up period for eight years.

REFERENCES