

## EXTRA-ADRENAL MYELOLIPOMA: A case report and review of the literature

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### ABSTRACT

Myelolipoma of the adrenal gland is a rare, benign and biochemically inactive tumor comprising mature fat and hematopoietic elements. Most of these lesions are discovered incidentally. The vast majority occurs within the adrenal glands, but extra-adrenal myelolipomas have also been reported. The typical extra-adrenal myelolipoma is a solitary, well-defined mass with in abdomen. We present a 70 year old male who came to us with pain of 5 months duration. Abdominal ultrasound examination showed a 12 x 8.5cm hyperechoic heterogenous semi-solid mass localized in the right-side. Histological examination of the mass revealed the features of extra-adrenal myelolipoma.

**KEY WORDS:** Myelolipoma, Extra-adrenal myelolipoma, Adrenal tumors.

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### INTRODUCTION

Myelolipoma was first described in 1905 by Gierke.<sup>1</sup> Adrenal myelolipoma is rare benign tumor consisting histologically of an admixture of adipose tissue and extramedullary hemopoetic elements within the adrenal glands.<sup>2</sup> Extra-adrenal perirenal myelolipomas are rare benign tumors that consist of adipose and hematopoietic tissue.<sup>3</sup> Extra-adrenal myelolipomas are single and usually occur within the abdomen. Its cause is still not clear.<sup>4</sup> In this report, we report a case of extra-adrenal myelolipoma in the light of recent literature.

### CASE REPORT

A 70-year-old man was admitted to Department of Urology with right side pain and swelling associated with fever. Physical examina-

tion revealed a mass palpated at the lower right quadrants. Ultrasound examination of the mass showed a 12 x 8.5cm hyperechoic semi-solid heterogeneous mass lesion, whose origin extending to abdomen and localized in the liver and right the kidney was not determined (Fig-1). Plasma and urinary hormonal studies were not helpful in diagnoses. Abdominal surgical exploration was performed on the intra abdominal mass. The specimen was referred to our department for evaluation.

Grossly, the mass was solitary lobulated covered by smooth capsule and some areas had hemorrhagic foci. The tumor measured 17 x



Fig-1: Ultrasound shows hyperechoic semi-solid heterogeneous mass lesion.

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Fig-2: Grossly, giant adrenal myelolipoma mass.

10 x 5cm and weighed 345g. The tumor was fairly well encapsulated by a thin layer of connective tissue (Fig-2). The cut surface had orange-yellow appearance, small cysts and areas of hemorrhage. In histologic examination the tumor was composed of mature adipose and hematopoietic tissues (Fig-3,4). Adrenal cortical tests were not present in relationship to our lesion. These findings were diagnosed as the right extra-adrenal myelolipoma.

### DISCUSSION

Myelolipoma is a rare, benign and biochemically non-functioning tumor composed of a variable amount of fat and myeloid tissue similar to that found in normal bone marrow. Extra-adrenal myelolipomas may have a predominance of either the hematopoietic or fatty component. Extra-adrenal myelolipoma should be morphologically differentiated from mass-

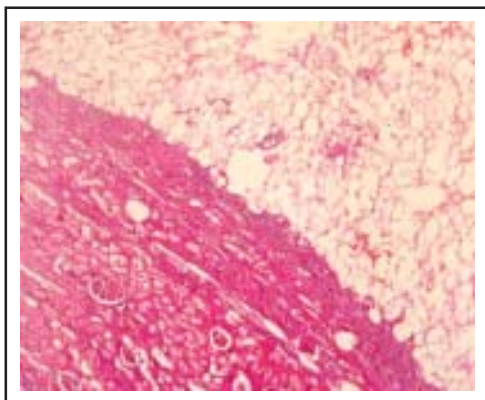


Fig-3: Renal parenchyma, adipose tissue and hematopoietic cells (HEX100).

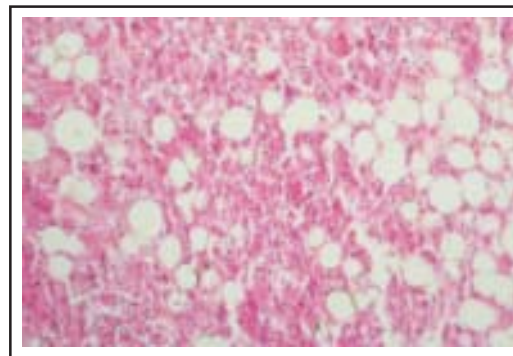


Fig-4A: Mature adipose tissue with hematopoietic cells (HEX200).

forming extra medullary hematopoiesis. The former is encapsulated or well-circumscribed, is composed of fat cells and has normal marrow hematopoietic elements. The latter lacks circumscription, and fat is not an integral component of the process.<sup>5</sup>

The occurrence in extra-adrenal sites, however, is quite rare. From autopsy studies, the incidence has been estimated as 0.4% in the general population. Approximately 36 extra-adrenal myelolipomas have been described in English literature.<sup>3</sup>

Pathogenesis of adrenal myelolipoma remains unclear. Theories include autonomous proliferation of bone marrow cells transferred during embryogenesis, degeneration of epithelial tissue of the adrenal cortex and metaplasia of mesenchymal cells.<sup>6</sup> Less frequently, they are believed to arise from adrenal cortical rests. A similar causative mechanism for extra-adrenal myelolipomas also seems likely.<sup>7</sup>

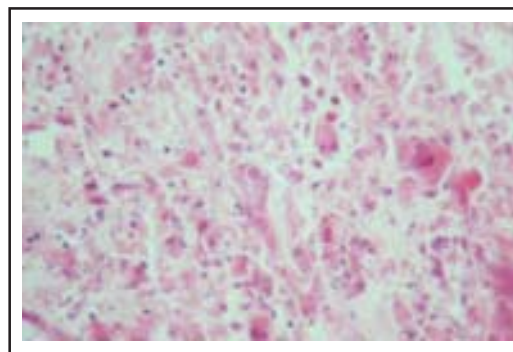


Fig-4B: High magnification of lesion. A cluster of megakaryocytes, scattered lymphocytes, and plasma cells (HEX400).

Myelolipoma is usually localized in the region of the adrenal gland. Extra-adrenal myelolipoma may occur in the retroperitoneum, thorax, pelvis, stomach, liver, lung, presacral area and in 3% of cases even in the mediastinum.<sup>2,9</sup> Extra-adrenal myelolipoma is solitary, well-defined mass within the abdomen.<sup>7,9</sup>

In extra-adrenal locations, radiologic evaluation, although helpful, may not be as definitive. Several fat-containing lesions, such as angiomyolipoma, liposarcoma, or lipomatosis, are more frequently encountered in extra-adrenal sites, kidney, or perirenal fat.

In conclusion, extra-adrenal myelolipomas, particularly those arising as perirenal masses, are rare. The differential diagnosis of a renal hilar neoplasm should include myelolipoma, although these lesions are rare. Since most of the results of histological examinations or autopsies indicate incidental findings, extra-adrenal gland myelolipoma should be considered clinically, radiologically and pathologically in differential diagnosis of adrenal gland lesions.

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