

GIANT MYELOLIPOMA OF THE ADRENAL GLAND

Ziaaddin Madani Kermani*, MD

** Associate Professor, Tehran University of Medical Sciences, Faculty of Medicine, Amir Alam Hospital:
Department of Pathology*

SUMMARY

Myelolipoma, a rare benign non-functioning neoplasm, is composed of mature adipose tissue and bone marrow elements. Its most common location is adrenal gland; however, extra-adrenal cases have been also reported. It is found in only 0.2% of all autopsies; 96% of the reported cases were detected on postmortem examination. The surgical symptomatic cases having been reported so far are few. In this paper, one case of myelolipoma with clinical presentation will be described and its clinicopathological features, pathogenesis, associated diseases, and diagnostic techniques will be discussed. It must be emphasised that correct diagnosis largely depends on the clinicians and surgeons' awareness of this rare and unique entity. In short, the diagnostic features of myelolipoma include: negative biochemical findings; radiolucency on routine x-ray film; a solid mass in ultrasonography; and, typical patterns on CAT magnifying resonance imaging (MRI).

INTRODUCTION

Myelolipoma, a non-functioning neoplasm of adrenal, is composed of mature adipose tissue and bone marrow elements. Extra adrenal locations are pelvic region and retroperitoneum.

Most of the reported cases were small and detected on postmortem examination of the adrenals; however, surgical cases have been recorded in the literature occasionally.

Our case was a surgical one with extraordinary features.

The presence of bone marrow elements in the adrenal gland was first described by Arnold in 1886 in the German literature and by Wooly in 1914 in the English literature. Gierke was the first to describe a detailed histological pattern of this tumor in 1905; Oberling suggested the term *myelolipoma* for this unique tumor.

The purpose of this paper is to report an unusual case of adrenal myelolipoma and discuss its clinicopathological characteristics.

CASE REPORT

A.N. was a 32-year-old white hermaphroditic patient who was admitted to the hospital with left lower quadrant pain and sense of heaviness, he denied other clinical symptoms relating to the tumor. On physical examination, he was found to have two small atrophic testes and small underdeveloped penis. X-ray revealed a large translucent mass in the left renal region. The patient was operated according to the preoperative diagnosis of renal cell carcinoma.

MATERIALS AND METHODS

GROSS CHARACTERISTICS

The specimen consisted of a large rather rounded greyish-yellow mass which measured 14x12x10 cm and weighed 1050 grams. Cut surface revealed a greasy variegated appearance composing of solid greyish-yellow areas and

dark-red soft hemorrhagic foci.

FIXATION AND PROCESSING

Thin two-centimeter sections were obtained and put in 10% formaldehyde solution for 24 hours for proper fixation, and then were placed in tissue processor for further fixation, dehydration and paraffin coating prepared paraffin blocks were cut with microtome and mounted on microscopic slides. Ordinary hematoxylin and eosin staining technique were used for preparation of final microscopic slides.

MICROSCOPIC PATTERN

Light microscopic sections revealed that adrenal cortical tissue composed of large cells having abundant clear cytoplasm and rather round small nuclei. There were also mature adipose tissue with clear cells and peripheral nuclei with aggregates of typical hematopoietic cells, Upon high magnification, all marrow elements were found to be composed of myeloid, erythroid, and megakaryocytic with orderly maturation. There were few scattered lymphocytes and plasma cells as well.

DISCUSSION

Myelolipoma is a benign non-functioning neoplasm which occurs with greater frequency in the adrenal glands; however, extra adrenal locations have been also reported in retroperitoneal and pelvic regions and recently

in intracranium. Most of the recorded cases were small and detected on postmortem examination, and only a few large and symptomatic cases have been reported in the literature, none of them were diagnosed preoperatively.

The pathogenesis of myelolipoma is obscure; although the condition is more likely to represent a hamartomatous than a true neoplasm and may be due to prolonged stress and stimulation with adrenocorticotrophic hormone. This tumor occurs most commonly in middle-aged or elderly individuals. In a large documented series, the age range was from 17-93 years. There was no sex preference. The incidence of this tumor upon postmortem examination of 2000 autopsies was 0.2%. Associated endocrine disturbances as Plaut noted were Addison's disease, Cushing's disease, hermaphroditism, obesity, virilism, and multiple endocrinopathy in 9 of 100 patients with myelolipoma. However, endocrine function of this tumor has never been demonstrated.

There is a single report in the literature in which myelolipoma was associated with nephrotic syndrome, this previously unreported clinical presentation of myelolipoma could have been caused by some substance secreted by the tumor, or could have been due to the compression of the renal vein. Although the pathogenesis of the nephrotic syndrome was not found in this patient, prompt postoperative

cessation of proteinuria and disappearance of edema was the clinical evidence of a cause and effect relationship between myelolipoma and nephrotic syndrome (1).

Usual clinical symptoms in surgical cases which have been reported so far were: pain, jaundice; microscopic hematuria; weakness and dyspnea on exertion; and, headache in the case of posterior fossa myelolipoma of intracranium (2). This tumor does not generally cause symptoms until it becomes large enough. Some clinical symptoms are related to hemorrhage (3).

Correct preoperative diagnosis is based largely on the surgeon's awareness of this rare benign tumor and proper usage of the current techniques such as: x-ray, angiography, IVP (Intravenous Pyelography) and sonography, and, the most important, interpretation of findings (3).

Our case exhibited several unusual features which make it somewhat unique among the previously reported cases. These uncommon features are summarised as follows:

1) Our case was a symptomatic surgical one which comprises only 4% of the total cases of myelolipoma (up till now the reported surgical cases are about 20);

2) the size of the specimen was considerably large, the first largest reported case was 1100 grams, described by Dyckman and Freedman in 1957;

3) low patient's age (32 years);

4) its association with hermaphroditism, a rare occurrence.

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