ISSN 2091-2889 (Online)
ISSN 2091-2412 (Print)

Journal of Chitwan Medical College 2015; 5(12): 46-49
Available online at: www.jcmc.cmc.edu.np

CASE REPORT

HUGE ADRENAL MYELOLIPOMA: A RARE ENTITY

G Pandey[1]*, B Thakur[2], M Devkota[2]

[1] Department of Pathology, B.P. Koirala Memorial Cancer Hospital.
[2] Department of Surgery, B.P. Koirala Memorial Cancer Hospital.

***Correspondence to:** Dr Greta Pandey, Department of Pathology, B.P. Koirala Memorial Cancer Hospital. E-mail: pandeygreta@hotmail.com*

ABSTRACT

Adrenal myelolipoma is a benign uncommon tumor of adrenal gland which is mostly diagnosed incidentally. Huge adrenal myelolipoma are very rare. Histopathological examination gives the confirmed diagnosis. Here we present a 76 years old female with huge retroperitoneal mass, diagnosed as adrenal myelolipoma on the basis of histopathological examinations. Surgical management is the treatment of choice for large symptomatic tumors.

Key words: *Adrenal, Hematopoetic elements, Myelolipoma, Retroperitoneal.*

DOI: http://dx.doi.org/10.3126/jcmc.v5i2.13157

INTRODUCTION

The term 'myelolipoma' was coined by Oberling in 1926; long after its description in 1905 by Gierke [1].These are rare tumors, detected incidentally during autopsy procedures with frequency of detection between 0.08-0.2% [2]. Primary adrenal gland myelolipoma accounts for 2.5 % of all adrenal tumors [3]. However in recent years with the use of USG, CT and MRI the incidental detection has increased [4].The majority of these lesions are small and asymptomatic. Some large symptomatic lesions have also been reported [5]. These are non-functioning tumors but have been reported to co-exist with other hormonally active tumors of adrenal gland [6].

There is equal sex predisposition with 5th to 7th decade being common age group [7].Grossly these are well defined tumors. On cut sections there are yellow to brown areas. Histopathological examination shows mixture of adipocytic tissue along with hematopoietic elements [3].No potential of malignancy for adrenal myelolipoma has been reported [5].

CASE REPORT

A 76 year old female patient presented with a history of pain abdomen for 3 years. There was progressive increase in pain along with distension of abdomen. Bowel and bladder habit was normal. Lab investigations show decreased hemoglobin level along with increased fasting glucose detected first time during pre operative investigation. Other investigations were within normal limit. CT scan revealed the presence of well defined retroperitoneal, predominantly fat containing lesion measuring 30x20cm posterior to liver and anterior to right kidney noted at anterior pararenal space. There is intralesional septations but no evidence of calcification noted. FNAC was also done, reported as benign lipocytic lesion. So taking into account, the size and location of tumor along with CT report, a provisional diagnosis of retroperitoneal liposarcoma was made.

With the clinical diagnosis of retroperitoneal liposarcoma, this patient underwent exploratory laparotomy. Operative finding was a solid bosselated mass 30x20x10 cm in the right side of retroperitoneum extending to RIF , diaphragm and to duodenum. There was displacement of right kidney and liver as

well. On Gross examination there was a well defined mass with smooth outer surface. Cut sections showed yellow smooth areas along with foci of hemorrhage (Figure 1).Cystic changes, myxoid changes or necrosis were not present. Multiple histological sections revealed mixture of mature adipose tissue along with hematopoietic elements without cellular atypia (Figure 2). These hematopoietic elements comprised of myeloid and lymphoid elements along with megakaryocytes .There was a thin rim of adrenal tissue as well (Figure 3).

Fig 1: Gross finding showing yellow fatty areas and hemorrhagic areas.

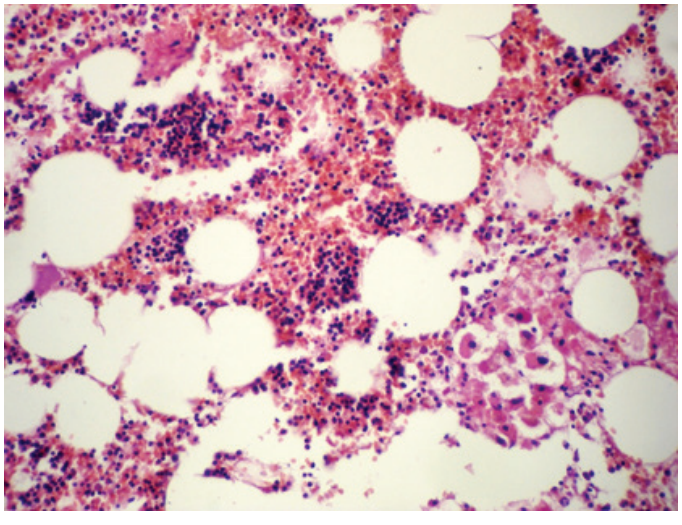

Fig 2: Histopathological picture (40 X)showing mature adipocytes, along with hematopoeitic elements

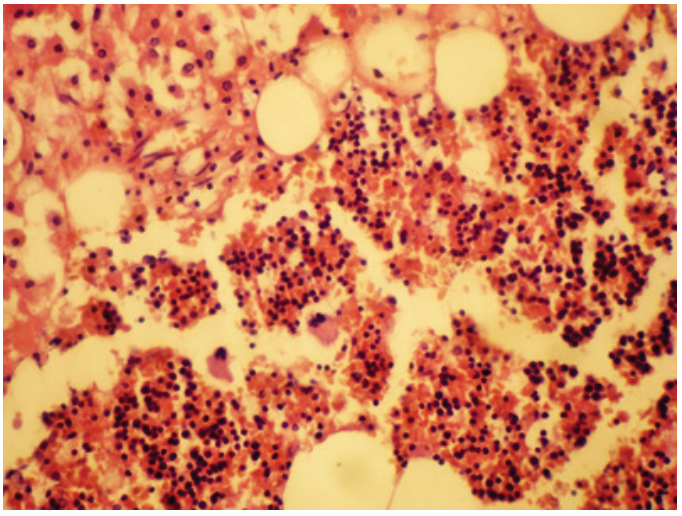

Fig 3: High power view(40X)showing adrenal tissue(upper left)and hematopoeitic elements including megakaryocytes(megakaryocytes).

DISCUSSION

Adrenal myelolipoma are rare entity in surgical practice. These lesions are characterized by presence within the adrenal gland of adult fat containing active bone marrow elements, with widely variable cellularity [8].The trilineage marrow components which include the myeloid, lymphoid and megakaryocytes are found [9].

They are called incidentalomas because most are diagnosed after autopsy or imaging procedures performed for some other diseases. Incidence rate ranges from 0.08% to 0.4% and less than 300 cases are found in literature before 2000 [10]. Myelolipomas are usually located in adrenal gland but occasionally they may be located at extra-adrenal sites like retroperitoneal presacral region, perirenal, renal hilum, spleen, lung and mediastinum [11].

The pathogenesis of adrenal myelolipoma is controversial. The suggested mechanisms include embryonic mesenchymal rests in adrenal glands, hematogenously seeded bone marrow emboli and metaplasia of reticuloendothelail cells as a result of chronic stress [6].A widely accepted theory is of adrenocortical cell metaplasia in response to stimuli like necrosis, inflammation, infection or stress. This chronic stimulus to adrenal gland which is evident more in advanced age could trigger the development of benign and malignant tumors as well as explains its incidence in older age group. The condition is

often associated with Cushing disease, obesity, hypertension and diabetes mellitus which can be characterized by major adrenal stimuli [7].

Clinically these patients present with flank pain and abdominal mass. The mechanical compression by larger tumors, tumor necrosis and retroperitoneal hemorrhage are reasons for painful sensations [6]. Hormone levels should be obtained, however, because co-existing functional cortical adenoma and myelolipoma have been reported. There are few case reports documenting presentations with virilisation, hypertension, hyperaldosteronism and hypercortisolism indicating hormone secretion leading to as well as pheochromocytoma Malignancy has not been reported in such lesions even when they attain large size [11].The radiological examinations including USG, CT and MRI are all effective for diagnosing adrenal myelolipoma in 90% of cases. The typical features include identification of fat, with CT scan being the most sensitive [7].

Grossly, they vary in size from microscopic to those filling the abdomen. They are not encapsulated but circumscribed lesion. On cut sections they are bright yellow with foci of tan brown discoloration [12]. This appearance depends upon the relative proportion of myeloid and fat component [13].Microscopic examinations show that they are composed of variable proportions of mature adipose tissue and active hematopoietic elements which comprises of lymphoid, myeloid and erythroid precursors as well as megakaryocytes. Areas of necrosis, hemorrhage, cysts formation and calcification are also evident in larger tumors. Myelolipomatous foci can be found within other pathological lesions such as adrenal hyperplasia or adenoma. In contrast to other extramedullary foci of hematopoeisis in adults associated with hematolytic disease, adrenal myelolipoma is practically always accompanied by normal bone marrow. The hematopoietic tissue contained erythroid , granulocytic cell lineage and few lymphoid cells along with megakaryocytes. A thin rim of normal adrenal tissue is also seen in sections [1,2,3,11,12].

Conservative management is recommended for asymptomatic tumors. Larger, symptomatic tumors are treated surgically [6].

CONCLUSION

Myelolipoma are rare tumors, huge myelolipoma are even rarer. Many times they are confused with liposarcoma as in our case. But earlier recognition is important as they have the risk of spontaneous retroperitoneal hemorrhage. Thorough preoperative biochemical study is mandatory to look for endocrine abnormalities. In previously unsuspected cases, once histopathological reports are available, endocrine abnormalities should be sought.

REFERENCES

1. Ersoy E, Ozdogan M, Demirag A, Aktimur R, Kulacoglu H, Kulacoglu S et al. Giant adrenal myelolipoma associated with small bowel leiomyosarcoma: A case report. *The Turkish journal of gastroenterology* 2006; 17(2): 126-129.

2. Olsson CA, Krane RJ, Klugo RC, Selikowitz SM. Adrenal myelolipoma. *Surgery* 1973 May; 73(5):665-70.

3. Lioyd RV, Kawashima A, Tishler AS.Adrenal soft tissue and germ cell tumours. In: *Pathology and Genetics: tumours of endocrine organs*. France: IARC press 2004;p169.

4. Daneshmand S, Quek ML. Adrenal Myelolipoma: Diagnosis and Management. *Journal of Urology* 2006;3(2):71-74.

5. Bandar AH, Muhammad MR, Amal HA, Mohammad AZ, Wafa AS. Adrenal myelolipoma a rare benign tumour managed laparoscopically: Report of two cases. *Journal of Minimal Access Surgery* 2009;5(4):118-120.

6. Hung JS, Huang KC. A huge adrenal myelolipoma. *Journal of Taiwan Urological Association* 2007;18(2):102-104.

7. Junaid N,Danish R,Fatema N.A, Ghulam N.S. Incidental detections of adrenal myelolipoma: A case report and review of literature. *Case reports in urology* 2013; 2013(article ID 789481): 3. http://www.hindawi.com (accessed 20 January 2013).

8. Rosai J. *Surgical Pathology*.10th ed.India: Elsevier: 2012.

9. Bishop E, Eble JN, Chen L et al. Adrenal myelolipoma show non random X-chromosome inactivation in hematopoeitic elements and fat: support for clonal origin of myelolipomas. *Am J Surg Pathol* 2006;30(7):838-843.

10. Settakorn J, Sirivanichai C, Rangdaeng S, Chaiwun B. Fine needle aspiration cytology of adrenal myelolipoma: case report and review of the literature. *Diagnostic Cytopathology* 1999;21:409-412.

11. Ramakant P, Mukha R, Cherian JA et al. Do Giant Adrenal Myelolipomas Behave Differently? A Clinicopathological Comparative Study. *World Journal of Endocrine Surgery* 2013;5(1):12-15.

12. Ernest EL. Tumors of adrenal gland. In: Fletcher DMC (ed.) *Diagnostic histopathology of tumors*.3rd ed. Philadelphia, PA: Churchill Livingstone, Elsevier. 2007;115-17.

13. Fletcher DMC. *Diagnostic Histopathology of Tumors*, 3rd ed. Philadelphia, PA: Churchill Livingstone Elsevier; 2007.