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Giant adrenal myelolipoma associated with small bowel leiomyosarcoma: A case report

İnce barsak leiomyosarkomu ile birlikte görülen dev adrenal myelolipom: Olgu sunumu

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Adrenal myelolipoma is a rare, benign and biochemically inactive tumor. It is usually diagnosed incidentally by radiological methods and is known to be associated with obesity, hypertension, endocrinological disorders and some malignancies. We report herein the association of a myelolipoma with a gastrointestinal stromal tumor. To our knowledge this is the first report of such an association to date. A 67-year-old male patient admitted to our clinic with abdomimal pain and fever; he had a history of hypertension and diabetes mellitus. In physical examination, a mass involving the right quadrants was palpated. Computerized tomography revealed a right retroperitoneal mass, probably originating from the kidney or cecum. In laparotomy, the tumor (12 cm radius and 1500 g) localized on the superior of right kidney was excised. Abdomen exploration revealed another mass with 10 cm radius 100 cm distal to the ligamentum of Treitz and segmental jejunal resection and anastomosis were applied. The pathological diagnosis was reported as myelolipoma for the retroperitoneal mass and leiomyosarcoma for the jejunal mass. Myelolipoma is a benign tumor, involving mature fat and hematopoietic stem cells. Pathogenesis is still not clear and the microscopical characteristics are hematopoietic, lipoid, and reticuloid cells and megakaryocytes. Myelolipomas are reported to be associated with some other malignancies (especially renal), but this is the first report showing the association with a leiomyosarcoma. Therefore, leiomyosarcoma should also be one of the possible associations kept in mind by the physician in the diagnosis and treatment of myelolipomas.

Key words: Adrenal myelolipoma, small bowel leimyosarcoma

Adrenal myelolipom nadir, benign ve biyokimyasal olarak inaktif bir tümördür. Çoğunlukla rastlantısal olarak radyolojik çalışmalar sırasında saptanır. Adrenal myelolipom, obezite, hipertansiyon, endokrin bozukluklar ve malign tümörlerle birliktelik gösterebilmektedir. Bu olgu sunumunda, adrenal myelolipom'nin gastrointestinal stromal tümör ile birlikteliği ilk kez bildirilmiştir. 67 yaşında erkek hasta üşüme titreme ateş ve karın ağrısı ile kliniğimize başvurdu. Öyküsünde hipertansiyon ve diyabet mevcuttu. Fizik muayenede karın sağ alt/üst kadranlarda kitle palpe edildi. BT'de sağ retroperitoneal bölgeyi işgal eden, böbreği aşağıya itmiş ve çekumdan köken almış muhtemel liposarkom veya çekum tümörü rapor edildi. Laparotomide, sağ retroperitoneal bölgede, böbrek süperiorunda yerleşmiş 12cm çapında, 1500gr ağırlığında kapsüllü kitle ve ince barsakta da Treitz ligamanından yaklaşık 100cm distalde 10cm çapında kitleler eksize edildi. Patolojik tanı adrenal myelolipom ve jejunal leimyosarkom olarak rapor edildi. Adrenal myelolipom, mature yağ ve hematopoetik kök hücrelerinden oluşmuş iyi huylu bir tümördür. Mikroskopik olarak adrenal myelolipom yağ dokusu içerisinde hematopoetik, lipoid, retiküloid hücreler ve megakaryositlerin görülmesiyle karakterizedir. Adrenal myelolipomun malignitelerle (özellikle renal karsinom) birlikteliği bildirilmiş olmakla beraber literatürde leimyosarkom ile birlikteliği bildirilmemiştir. Adrenal myelolipom-Sarkom birlikteliğinin ortak patojenezi konusunda da yeterli bilgi bulunmamaktadır. Klinisyenler adrenal myelolipom tanı ve tedavisini planlarken olası patolojik birliktelikler arasında leimyosarkomu da gözönüne almalıdır.

Anahtar kelimeler: Adrenal myelolipom, ince barsak leimyosarkomu

INTRODUCTION

Myelolipoma is a lesion characterized by presence within the adrenal gland of adult fat containing active bone marrow elements. It was first described by Gierke in 1905 and then by Oberling in 1929 (1). Most cases are found incidentally, either at autopsy (0.4%) or through computerized tomography (CT) scanning done for other reasons. Only occasionally will the lesion attain a size large eno-

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ugh to become clinically apparent. It is usually unilateral and hormonally inactive, and most of the patients are obese adults. It is primarily diagnosed in the fifth to seventh decades of life (1-3). Myelolipoma has been seen in association with adrenal cortical tumors accompanied by Cushing's syndrome and with congenital adrenal hyperplasia. Some associations with endocrinological pathologies or malignancies have also been reported for myelolipoma. In the English literature, there are a few reports on large series of patients with adrenal myelolipomas. In this report, a case of adrenal myelolipoma associated with a jejunal leiomyocarcinoma is presented.

CASE REPORT

A 67-year-old man was admitted to the Department of General Surgery with abdominal pain associated with fever and recently diagnosed high blood glucose level. He had no significant history except for hypertension. Physical examination revealed a mass palpated at the upper and lower right quadrants which was slightly painful. CT showed a mass of 12 x 10 cm between the right lobe of the liver, right kidney and the ascending colon. The mass had air levels inside and was interpreted as a retroperitoneal sarcomateous lesion that had fistulized to the cecum or as a cecal mass dislocating the kidney. Surgery was planned to explore the mass.

Abdominal surgical exploration was performed in order to explore the intraabdominal mass. A huge capsulated tumor 12 cm in radius and weighing 1500 g was removed from the right retroperitoneal area (Figure 1). A tumor of 10 cm located 100 cm distal from the ligamentum of Treitz was found incidentally and resected segmentally (Figure 2). No significant morbidity was reported after the operation. The histopathological diagnosis of the retroperitoneal tumor was myelolipoma, and the tumor from the jejunum was reported as gastrointestinal stromal tumor (leiomyosarcoma). The patient was followed for 16 months with no significant problems. In the 16th month of the follow-up, two masses of the liver were reported in the routine control CT, which were thought to be metastases of the jejunal sarcoma.

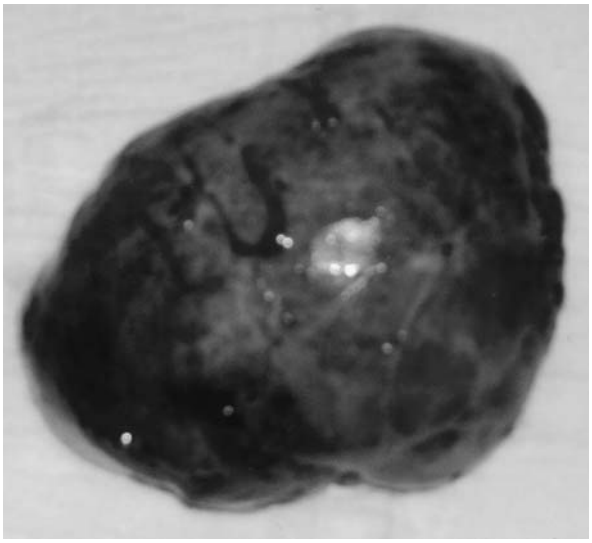

Figure 1. Myelolipoma specimen

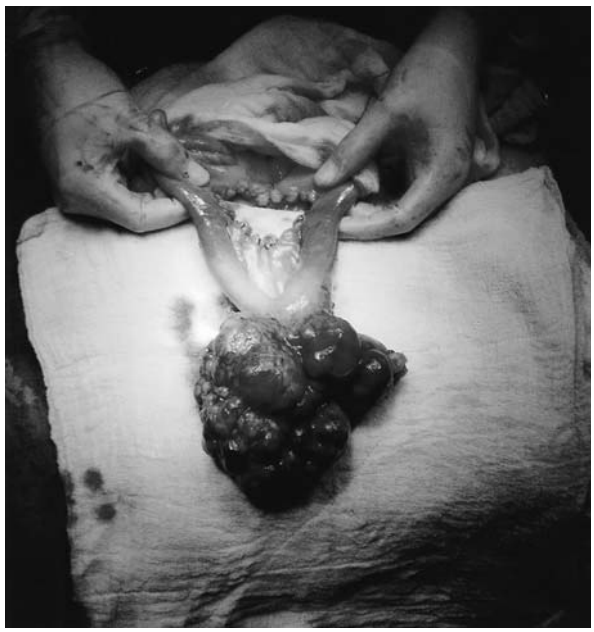

Figure 2. Intraoperative image of the jejunal leiomyosarcoma

DISCUSSION

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. The most accepted theory is adrenocortical cell metaplasia of the reticuloendothelial cells of blood capillaries in response to stimuli such as necrosis, infection or stress (1). It is of interest that adrenal myelolipomas may be secondary to prolonged stress and excessive stimulation by adrenocorticotropic hormones and may be associated

with endocrine disorders such as hermaphrodism, Cushing's disease, Addison's disease, or obesity of uncertain etiology. Selye and Stone produced adrenal myelolipomas in rats by injection of crude adrenocorticotropic hormone and testosterone (4). Adrenal myelolipomas are composed of fat and hematopoietic elements, so the theoretical possibilities of pathogenesis include bone marrow embolization. Microscopically, adrenal myelolipoma consists of hematopoietic, lipoid and reticuloid cells in fatty areas, as well as myeloid precursors (megakaryocytes) (5).

Most adrenal myelolipomas are asymptomatic, diagnosed accidentally during imaging procedures, and they are usually less then 4 cm in diameter (6). However, some cases are large and are diagnosed by complications like compression of adjacent organs, rupture and hemorrhage or flank pain. In the literature, tumor size has ranged between 1 and 34 cm in diameter, with the heaviest weighing 6000 g (7, 8).

Differential diagnosis includes retroperitoneal lipoma, liposarcoma, exophytic renal angiomyolipoma, adrenal adenoma and primary or metastatic malignant adrenal tumors (3, 6). CT is a highly sensitive technique in the imaging and diagnosing of those tumors. If the tumor is huge as in our case, it should be removed surgically. On the other hand, a fine needle aspiration is indicated in small and tomographically indistinguishable retroperitoneal tumor. The incidence of calcification is 27% and it might be a feature of adrenal myelolipomas on imaging (6). In adrenal masses with the presence of fat, even the punctuate calcifications, should alert the radiologist for the diagnosis of mylolipoma (2,7). Ultrasonography usually reveals hyperechogenic tissue with propagation speed artifact, while CT will reveal lesions with the attenuation of adipose tissue. If doubt persists, especially with regard to possible malignancy, further information can be obtained by angiography, which will show a hypovascular tumor, magnetic resonance imaging, and tomography-guided fine needle aspiration biopsy for histological diagnosis (9). The endoscopic ultrasound-guided fine needle aspiration biopsy is a highly specific and safe technique for confirming the diagnosis of adrenal tumors (10).

Although myelolipoma is most common in adrenal glands, it may also be encountered as an isolated soft tissue mass, especially in the pelvic region, in patients without hepatosplenomegaly and without any evidence of hematopoietic disorders. It must be distinguished from extramedullary hematopoiesis associated with various myeloproliferative diseases myelosclerosis, severe anemia and skeletal disorders. In the recent literature, there are 36 cases reported in extra-adrenal regions. Among them, 14 cases were noted in the presacral soft tissue and the others elsewhere (3).

The association of adrenal myelolipoma with obesity, hypertension, atherosclerosis, diabetes mellitus and other chronic diseases has been described previously (4, 7, 10-13). Associations with some other malignancies were also previously described (14). Although we have knowledge regarding the associations of myelolipoma, it is not possible to postulate it physiopathologically because most of the diseases said to be associated with myelolipoma occur in the fifth to seventh decades of life, and the disease is usually diagnosed incidentally. Furthermore, we do not have exact data about the period between the onset and the diagnosis. However, no cases of myelolipoma and sarcoma were found in the literature. Our patient had hypertension and a recently diagnosed diabetes mellitus. High blood pressure is the most common accompanying disorder with myelolipoma, which interestingly has been noted to normalize after the removal of the tumor in some cases (10). It could be explained endocrinologically (disorders of the adrenal gland) or by the superior renal vasculature compression of a huge myelolipoma. It is not apparent how these medical conditions contribute to the etiology or the pathogenesis of the tumor. Although associations with endocrinological disorders have been reported in most of the articles about myelolipoma, the diagnosis does not rely on endocrine evaluation, since the tumor has been defined as being hormonally inactive. However, the association of myelolipoma and endocrine disorders accounts for 85%, and the mechanism is still unclear (4, 7).

Management of adrenal myelolipomas should be individualized. It has been recommended that surgery should be reserved for symptomatic tumors as well as asymptomatic tumors more than 4 cm in diameter, as they have a higher hemorrhage risk. Small tumors can be monitored radiologically with periodic ultrasonography or CT (15-17).

The association of myelolipoma and malignancies is also unclear thus far. The reports in the literature are all case reports, and so it is doubtful to claim about a pathogenetic association. Although a relationship between endocrinological pathologi-

es and myelolipoma has not been postulated, it is significant statistically. There are no reports in the recent literature about a myelolipoma and a jejunal leiomyosarcoma. Our case has the characteristic specifications of myelolipoma like accompanying hypertension, diabetes mellitus and the age of the patient, and also an accompanying malignancy.

Gastrointestinal stromal tumors are the most common mesenchymal tumors arising from the small intestine and comprise up to 15% of all small bowel malignancies. Gastrointestinal stromal tumors comprise the vast majority of tumors that were formerly classified as leiomyomas, leiomyosarcomas and smooth muscle tumors of the intestine. Jejunal leiomyosarcoma is a rare variety of malignant small bowel tumor and accounts for less than 15% of the malignant small bowel tumors. The current therapy for leiomyosarcoma is segmental intestinal resection. If the diagnosis is known prior to resection, wide lymphadenectomy can be avoided because gastrointestinal stromal tumors are rarely associated with lymph node metastasis. An aggressive management approach using a combination of surgery and a newer technique like radiofrequency ablation can be attempted in a subset of patients with limited metastatic spread to liver to prolong the long-term survival (15). Chemotherapy or radiotherapy usually gives very poor results in leiomyosarcoma management. This also complicates the management of metastatic disease.

Although most myelolipomas are clinically silent and benign tumors, the clinician should be aware of their presentation to avoid unnecessary surgical interventions. The increasing number of cases reporting the association of myelolipomas with various endocrine disorders and malignancies emphasizes the importance of a rational and cost-effective evaluation and treatment method. The subject still remains unclear pathogenetically and needs to be investigated more. To our knowledge, this report is the first about the association of a leimyosarcoma and myelolipoma. Although it cannot be said with certainty whether or not this was a coincidental association, we can say that a clinician should also be aware of possible associated gastrointestinal malignancies while managing the patient with myelolipoma.

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