Adrenal myelolipoma and primary hyperparathyroidism due to ectopic parathyroid adenoma: case report and review of the literature

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ABSTRACT

Background: Adrenal myelolipoma is a rare benign neoplasm which is composed of mature adipose tissue and scattered islands of hematopoietic elements. Usually, myelolipoma is small (<4 cm), asymptomatic, and nonsecreting. In some cases, it may cause symptoms such as chronic pain. It can also be hormonally active, as there is a relatively high incidence (10%) of associated endocrine disorders, such as Cushing's syndrome, Conn's syndrome, congenital adrenal hyperplasia, adrenal insufficiency, and pheochromocytoma. On the other hand, primary hyperparathyroidism is a common disorder caused by over-activation of parathyroid glands, resulting in the excessive release of the parathyroid hormone.

Case Presentation: Herein, we present a female patient who presented with abdominal pain due to adrenal myelolipoma, who was also found to have primary hyperparathyroidism and we discuss the possibility of the coexistence of multiple endocrine neoplasia type 1 syndrome.

Conclusion: To our knowledge, this is the first case of a coexistence of an adrenal myelolipoma together with an ectopic parathyroid tissue adenoma. The combination of an adrenal myelolipoma with an ectopic parathyroid tissue adenoma has never been described before and may or not be just a coincidence.

Keywords: Adrenal myelolipoma, primary hyperparathyroidism.

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Background

Adrenal myelolipoma is a rare, usually small, and asymptomatic benign neoplasm, which is located in the adrenal gland and comprises mature adipose tissue and intermixed myeloid tissue. Although usually small and asymptomatic, in some cases, these tumors are hormonally active with Conn's disease, Cushing's disease, and pheochromocytoma being some of the clinical manifestations. Furthermore, adrenal myelolipomas can present with symptoms, such as chronic pain and hemorrhage, and in these patients surgical resection is the treatment of choice [1,2]. Primary hyperparathyroidism is the third most common endocrine disorder after diabetes and thyroid disease and is characterized by the autonomous production of parathyroid hormone (PTH). The most common clinical presentation is asymptomatic hypercalcemia, although in some cases serum calcium is normal (normocalcemic primary hyperparathyroidism). The vast majority of cases are sporadic and primary hyperparathyroidism is classified as being caused by single adenoma, hyperplasia, multiple adenomas, and carcinoma. Less frequently, an ectopic parathyroid adenoma is responsible, the occurrence of which is uncommon, presenting 3%-4% of all parathyroid adenomas. The vast majority of them are located in the anterior mediastinum near the thymus and surgical treatment remains the gold standard in these patients. Familiar parathyroid disorders are responsible of 5% of primary hyperparathyroidism and include multiple endocrine neoplasia type 1 (MEN1) and 2A, hyperparathyroidism jaw-tumor syndrome and isolated familial hyperparathyroidism [3].

Case Presentation

A 61-year-old woman came to our hospital with a short history of abdominal and flank pain. Her past medical history was remarkable for arterial hypertension for which she received treatment with amlodipine and valsartan. On physical examination, she had high blood pressure. Serum electrolytes including potassium were normal, while the rest laboratory testing showed hypercalcemia (Ca= 12.5 mg/dl), hypophosphatemia (p = 2.2 mg/dl), high alkaline phosphatase = 240 U/l, high PTH = 230 pg/ml, and low 25(OH) Vitamin D (6.78 ng/ml). 24-hour urinary calcium was raised (526 mg/24 hours) with Ca/Cr ratio = 0.02, a

finding which excluded follicular hormone. The patient was diagnosed with primary hyperparathyroidism. First-line imaging workup included neck ultrasonography, which showed a multinodular goiter with no definite evidence of parathyroid adenoma, while thyroid-stimulating hormone in serum was within the normal range. Then, a sestamibi scintigraphy was carried out and showed an ectopic parathyroid adenoma in the anterior mediastinum, a finding which was confirmed by magnetic resonance imaging (MRI) (Figures 1 and 2). Gastroscopy showed a peptic ulcer compatible with primary hyperparathyroidism, while the abdominal ultrasound revealed a myelolipoma (8 × 8 cm) of the right adrenal gland, which was confirmed by computer tomography (CT) and MRI (Figure 3). In order to determine the hormonal activity of these findings, we carried out deydroepiandrosterone sulfate, Δ4A, 17-OH-progesterone, testosterone, sex-hormone binding globulin, renin, and aldosterone, which were within the normal range. Furthermore, we investigated the possibility of existence of the MEN1 syndrome through specific laboratory tests (gastrin, CgA, vasoactive intestinal peptide, and pancreatic polypeptide), but these results were negative too. The patient underwent a surgery through which resection of ectopic adenoma was carried out, while she denied to be surgically treated for the adrenal myelolipoma. One year later, she remains in a good condition.

Discussion

Adrenal myelolipoma is a rare benign lesion, which consists of fatty and hematopoietic elements and is described as a mesenchymal and stromal tumor of the adrenal cortex [1]. Usually, myelolipoma is small, measuring less than 4 cm, and asymptomatic, until it reaches larger sizes, and then may become symptomatic with back and abdominal pain due to the increased pressure of the surrounding structures by the presence of the tumor. In addition, myelolipoma is usually hormonally inactive (90% of the cases), although there are some case reports that suggest their association with overproduction of adrenal hormones [1,2]. The recommended management in asymptomatic myelolipomas less than 4 cm is observation, under the condition that malignancy has been ruled out. In these cases, the patient undergoes radiological evaluation at 3 and 6 months and then annually for the next 2 years, while hormonal laboratory tests should be carried out annually for the next 5 years [1]. On the contrary, although myelolipoma is a benign tumor, in cases of symptomatic tumors, growing tumors, hormone producing tumors, and tumors measuring >6 cm, surgical excision is the treatment of choice in order to prevent major complications [1,2]. Primary hyperparathyroidism is characterized by abnormal regulation of PTH secretion by calcium, resulting in the hypersecretion of PTH relative to the calcium concentration in serum.

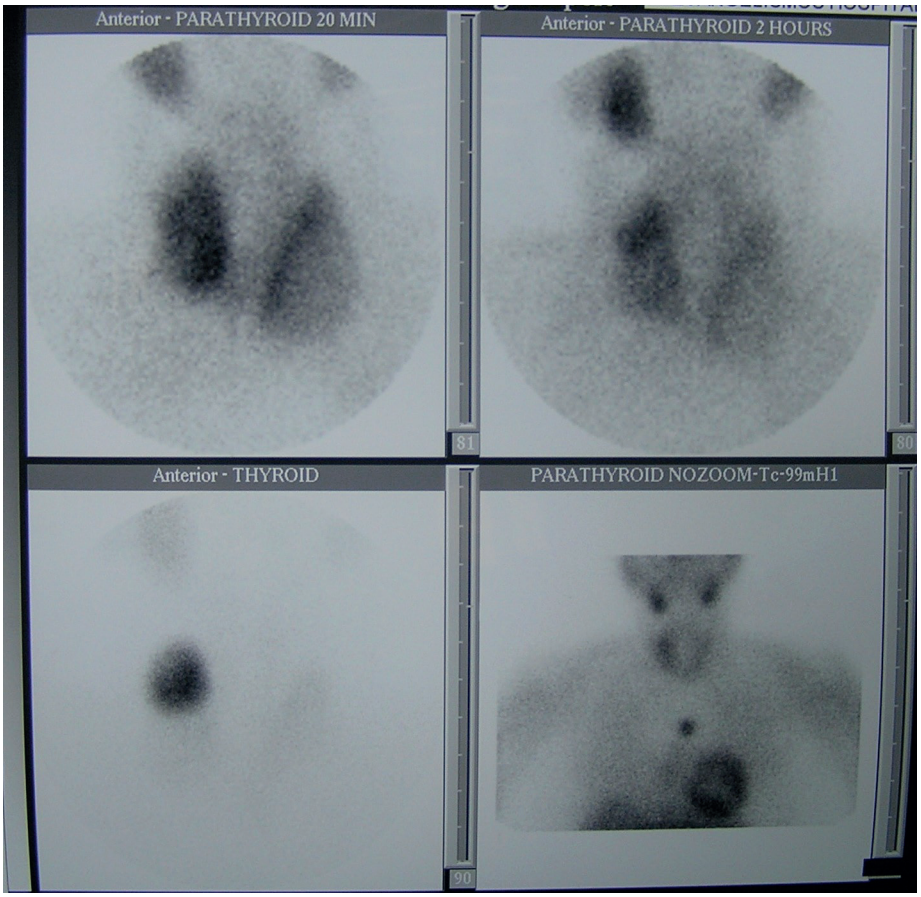

Figure 1. *Sestamibi scintigraphy.*

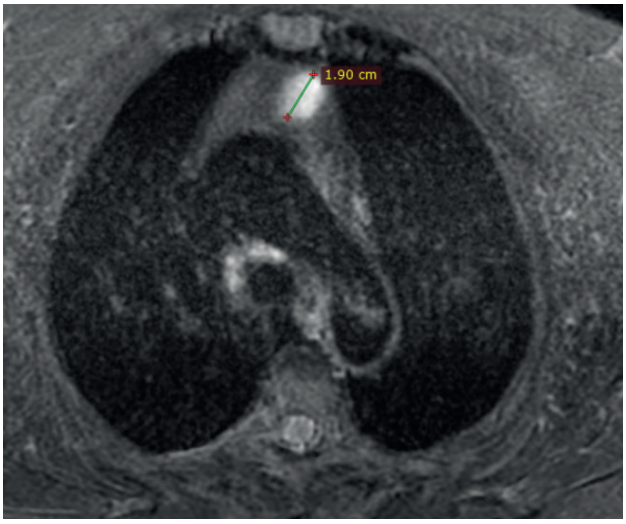

Figure 2. *MRI: ectopic parathyroid adenoma in the anterior mediastinum.*

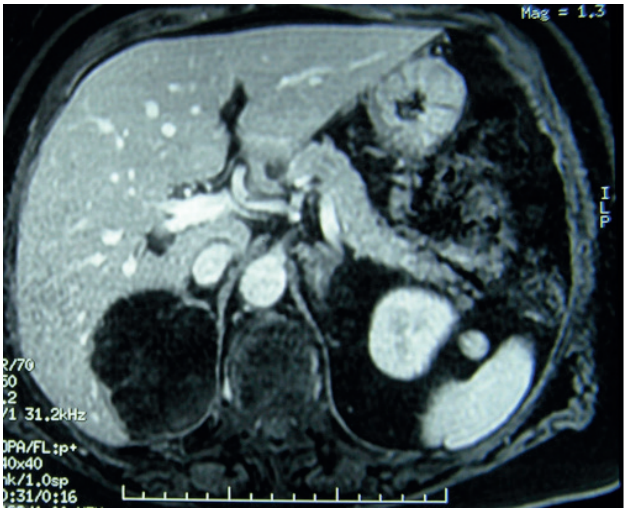

Figure 2. *MRI: Myelolipoma of the right adrenal gland.*

Common clinical presentations include joint pain, kidney stones, hypertension, osteopenia, and peptic ulcers. The main causes are head and neck radiation exposure, radioactive iodine therapy, calcium intake, and less frequent genetic abnormalities in the multiple endocrine neoplasia syndromes. Pathologic conditions that have been found in patients with hyperparathyroidism are adenoma, glandular hyperplasia, and carcinoma, with adenoma being the main cause in the vast majority of cases [3]. Neck is the most common location of parathyroid adenoma, while 0.3%-8% of the cases occurs in ectopic sites [3]. The clinical and laboratory findings in these cases seem to be more severe, when compared to those with eutopic adenomas, since the former may present with higher calcium levels in serum and more frequent primary hyperthyroidism-related bone disease [3]. Ectopic parathyroid adenomas are difficult to be detected by imaging modalities due to their small size, which is usually between 1 and 2 cm. Ultrasonography and sestamibi scintigraphy are used to diagnose parathyroid adenomas. CT and MRI have a higher sensitivity for localization of parathyroid adenomas compared to sestamibi scintigraphy and, furthermore, are more sensitive in detecting smaller lesions. Surgery resection is the treatment of choice carried out via median sternotomy and thoracotomy, although in some cases video-assisted thoracoscopic surgery seems to be preferred, as it has shown safety and fewer complications [3].

To our knowledge, this is the first case of a coexistence of an adrenal myelolipoma together with an ectopic parathyroid tissue adenoma. In 1999, Li-Fern et al. [4] had published the co-existence of myelolipoma with parathyroid tissue adenoma, while Saunders et al. [5] had recently reported the same coexistence.

Conclusion

Adrenal myelolipomas should be resected if symptomatic. The combination of an adrenal myelolipoma with an ectopic parathyroid tissue adenoma has never been described before and may or not be just a coincidence. Further studies are needed to confirm or not the coexistence of the above-mentioned clinical entities.

What is new?

Adrenal myelolipoma is a rare benign neoplasm which comprises mature adipose tissue and scattered islands of hematopoietic elements. On the other hand, primary hyperparathyroidism is a common disorder caused by over-activation of parathyroid glands, resulting in excessive release of PTH. Herein, we present a female patient who presented with abdominal pain due to adrenal myelolipoma, who was also found to have primary hyperparathyroidism, and we discuss the possibility of coexistence of MEN1 syndrome.

List of Abbreviations

CT	Computer tomography
MRI	Magnetic resonance imaging
PTH	Parathyroid hormone

Consent for publication

Written consent was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

Author details

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Summary of the case

1	**Patient (gender, age)**	She is a 61 year old female patient
2	**Final diagnosis**	An adrenal myelolipoma with co-existence of an ectopic parathyroid tissue adenoma
3	**Symptoms**	Abdominal and flank pain
4	**Medications**	amlodipine, valsartan
5	**Clinical procedure**	CY, MRI, Sestamibi scintigraphy
6	**Specialty**	Endocrinology