Case Report

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A Case Report of Sarcoma Near the Right of Adrenal Gland as an Rare and Important Differential Diagnosis of Adenoma in It on a 46-year-old Woman

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ABSTRACT

BACKGROUND AND OBJECTIVE: Retroperitoneal sarcoma constitutes 3-5% of all soft-tissue sarcomas and is often detected around 50 years of age. Soft-tissue sarcomas are categorized into four different types of well-differentiated, undifferentiated, myxoid and pleomorphic cells. Myxoid cells tend to cause lower-extremity involvement. In this study, we present a case of sarcoma of the right side of adrenal gland in a 46-year-old woman as a rare and important differential diagnosis of adenoma.

CASE REPORT: A 46-year-old woman referred to the Endocrine Clinic of Ayatollah Rohani Hospital of Babol to examine a 13 cm mass, which was randomly detected on the right side of adrenal gland via abdominal sonography. Sonography was requested due to flank pain, accompanied by night fever. Hormonal evaluation was negative and surgery was performed due to the large size of tumor mass. Pathological reports were indicative of a low-grade myxoid tumor. During four months of follow-up, the patient experienced no particular complications and underwent radiotherapy on a regular basis.

CONCLUSION: According to the results of this study, retroperitoneal masses can be posed in the rare differential diagnosis of adrenal tumors.

KEY WORDS: Retroperitoneal Mass, Adrenal Mass, Myxoid Tumor.

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Introduction

Retroperitoneal sarcoma is quite a rare condition, constituting only 3-5% of soft-tissue sarcomas (1). According to statistics, the annual incidence of retroperitoneal sarcoma is 2.7 cases per one million individuals in the world (2). These masses are usually detected in patients around 50 years of age; however, retroperitoneal sarcoma is known to occur within a wide age range. According to the literature, the prevalence of these masses is equal between men and women (3). Considering the morphological features and cytogenetic abnormalities, soft-tissue sarcomas are categorized into four main types of well-differentiated, undifferentiated, myxoid and pleomorphic cells (4). The anatomic distribution of tumors is associated with the histology of tissues; for instance, pleomorphic tumor cells, which are differentiated from myxoid cells, tend to cause lower-extremity involvement, while well-differentiated tumors mostly appear in the retroperitoneal space. Well-differentiated tumors often recur locally via distant metastases, and the five-year survival rate has been estimated to be over 90%. In addition, death due to the complications of surrounding critical organs usually occurs in the retroperitoneal space (5).

According to autopsy studies, the prevalence of adrenal incidentaloma is approximately 2%, covering a range of 1-9%. The prevalence of this tumor is reported to be higher in obese and diabetic individuals, as well as hypertensive patients (6).

When a mass is detected in the adrenal region, adrenal adenoma needs to be differentiated from other involved adjunct organs. In this paper, we present a case of low-grade myxoid tumor in the lower extremity of the right adrenal region with a difficult differential diagnosis of adrenal adenoma due to the proximity of the mass to the right side of the adrenal gland.

Case report

A 46-year-old woman referred to the Endocrine Clinic of Ayatollah Rohani Hospital in Babol to examine a 13 cm mass, which was detected randomly on the right side of the adrenal gland via abdominal sonography. The patient had complaints of weight loss and night fever over the past few months. She had been diagnosed with possible urinary tract infection, and antibiotics, which were prescribed for the fever and flank pain, were not effective. The evaluation of the patient was performed via laboratory tests and CT scan, without oral and intravenous contrasts.The obtained test results were as follows: normocytic anemia, ESR of 124H, and negative C-reactive protein (CRP) and purified protein derivative (PPD) tests. Moreover, urine sediments were inactive, and urinary catecholamines, metanephrines and vanillylmandelic acid were negative within 24 hours. In addition, low-dose dexamethasone suppression test, testosterone and dehydroepiandrosterone sulfate (DHEA-S) were negative.

Kidney-Urine-Bladder (KUB) Radiogrpah: During KUB radiography, a non-homogeneous mass with multiple foci of calcification was detected in the right adrenal gland (fig 1).

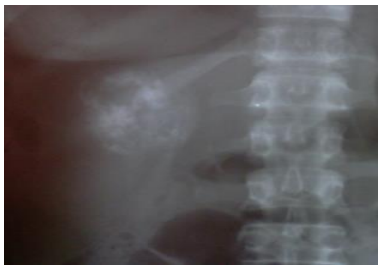
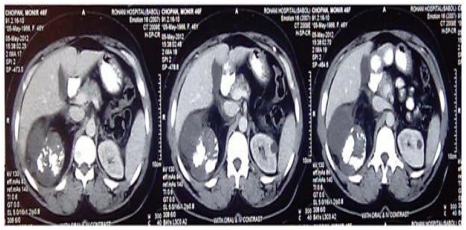


Figure 1. The image of a calcificated mass in the right adrenal area in KUB radiography

During the CT scan, a mass (diameter of 13 cm) with necrosis and calcification foci was detected in the right adrenal area, which had dropped the kidney downwards (Figure 2). Therefore, surgery was performed in order to remove the mass. In addition, a large mass, which was causing pressure on the adjacent tissues, was successfully removed. Necrosis and calcification were observed in the macroscopic cuts of the mass, and low-grade myxoid tumor cells were detected in the microscopic cuts.

A



B

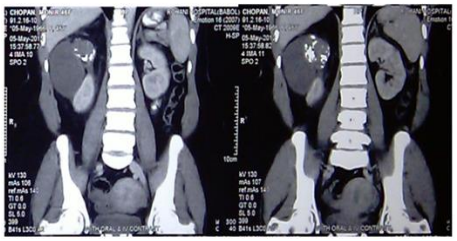


Figure 2. A) The calcified mass in the right adrenal area in the axial T1MRI section;
B) The same mass in the right adrenal area in the coronal T1MRI section

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Discussion

In the present study, we detected a mass on the right side of the adrenal gland in a middle-aged woman, which was initially assumed to have originated from the adrenal gland; however, after the surgical intervention, the origin of the mass was found to be retroperitoneal as identified by the pathology of liposarcoma with myxoid cells. According to a study by Gierke et al., the majority of such masses were asymptomatic and only a limited number of patients with these masses complained of flank pain. However, in the current study, the patient complained of fever and weight loss, in addition to flank pain. In another study, Fletcher CD categorized retroperitoneal sarcomas into four main subtypes of well-differentiated, undifferentiated, myxoid and pleomorphic cells (4).

In the present study, liposarcoma was detected by the type of myxoid cells during the pathological analysis of the mass. It seems that the anatomic distribution of lipoprotein sarcomas largely depends on tumor histology, as myxiod and pleomorphic cells tend to involve the adjacent organs, while well-differentiated tumors mostly occur in the retroperitoneal space.

On the other hand, well-differentiated tumors often recur locally and are associated with the lowest distant metastasis. The five-year survival rate in these tumors is 90% and deaths caused by the involvement of the adjacent, sensitive organs normally occur in the retroperitoneal space (5). Furthermore, differentiated tumors are associated with local complications; although systemic metastases may be observed in the lungs, metastasis is significantly lower (15-10%), compared to the predicted morphology of these tumor cells; therefore, the five-year survival rate is approximately 75% (7). Myxoid cell tumors represent a chain of morphologic rate (8). On the other hand, pleomorphic tumors are known to have a minimum prevalence rate; nevertheless, they tend to be aggressive with a high metastasis rate and a five-year survival rate of 30-50% (9). As for the treatment of retroperitoneal tumors, surgical excision is still considered as the most effective method (10). Large retroperitoneal tumors (>10 cm) have a high level of cell differentiation, which leads to difficulties in the diagnosis of retroperitoneal lipoma. In most cases, complete resection with a healthy margin size is a complicated process. Furthermore, radiation is often difficult due to the large size of tumors and the associated morbidity. In case of low-grade and well-differentiated tumors, adjuvant chemotherapy is considered as an effective approach. The use of adriamycin and combined chemotherapy with ifosfamide is also practical for various high-grade and differentiated tumors such as myxoid cells and pleomorphic tumors with a partial response of 50% among patients (11). In addition, a complete response to chemotherapy has been observed in less than 10% of patients.

In conclusion, the results of the present study indicated that retroperitoneal masses are extremely rare in the differential diagnosis of adrenal tumors.

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