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Case Report

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Adrenal involvement due to sarcoidosis presenting with Addison's disease

Fatih Acehan[1], Enes Seyda Şahiner[2]
[1]Department of Internal Medicine, Ankara City Hospital, Ankara, Turkey
[2]Department of Internal Medicine, Ankara City Hospital, Ankara, Turkey

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ABSTRACT

In this case report, we presented a case presenting with adrenal insufficiency, and which was thought to have sarcoidosis related involvement of adrenal gland in the etiology. A 68 years old female patient was diagnosed with sarcoidosis for having bilateral hilar lymphadenopathy, nodular lesions in the lungs, elevated serum angiotensin converting enzymes, and the colon and velum biopsies being concordant with noncaseified epitheloid granuloma. In addition, fatigue, weight loss, hyperpigmentation, hypotension, hyponatremia which had been continuing for four months until the admittance of the patient, suggested adrenal insufficiency. With the measurement of serum cortisol levels low and adrenocorticotropic hormone levels high in the further tests, the adrenal insufficiency diagnosis was established. After excluding other reasons of Addison's disease (autoimmune, tuberculosis, malignity, etc.), the adrenal insufficiency was thought to be related to sarcoidosis involvement.

Correspondence Address:Fatih Acehan Fatih Üniversiteler Mahallesi 1604. Cadde No: 9 Çankaya / Ankara - Türkiye / E-posta: acehanf@gmail.com

Introduction

Sarcoidosis is a chronic, systemic, and inflammatory disease characterized by noncaseified granulomas. While the reason of sarcoidosis is not known for certain, it is accepted that both genetic predisposition and environmental factors play a role. Sarcoidosis can involve all organs and systems; most frequently the lungs and the intrathoracic lymph nodes, and less frequently the skin, the eyes, and the skeletal system.[1] Rarely, it can involve the upper respiratory system and the endocrine and gastrointestinal organs.[2] There is a limited number of cases in the literature, in which the adrenal glands are involved and presenting with adrenal insufficiency. In this case report, a case presenting with adrenal insufficiency and which is thought to have sarcoidosis related involvement of the adrenal gland in the etiology, will be discussed.

Case Presentation

The 68 years old female patient had applied to the internal diseases outpatient clinic with nausea, throwing up, fatigue, weight loss, and constipation complaints. In the system query, it was found that the dizziness, fatigue, nausea, throwing up and constipation complaints had been continuing for four months, and it was learned that the patient had lost 8 kilos in the last four months and the skin color had darkened especially over the joints. In the physical examination, all vital signs were stable, except for the hypotension (90/50 mmHg). There was hyperpigmentation on the face, the neck, the dorsum of the hand and on the mucous membranes. There were not any pathologies in the examinations of other systems. In the laboratory tests, the c-reactive protein was found 32mg/dl (0-5), the erythrocyte sedimentation rate 72mm/h (0-20), Na:128mmol/l, K:5.41mmol/l, Ca:10.1mg/dl. There were not any pathologies in other laboratory tests. Hilar enlargement was present in the chest radiography. The electrocardiography had a normal sinus rhythm. The initial examinations were requested suspecting adrenal insufficiency considering the initial complaints and the laboratory tests. In the further examinations the serum cortisol

level was found low, 1.9 uq/dl (6.2- 19.4), and the Adrenocorticotropic Hormone (ACTH) level was found high, 1841pg/ml (7.2 - 63.3). The pituitary hormones, except for the ACTH, were found normal. The patient was diagnosed with adrenal insufficiency. Primary adrenal insufficiency was suspected due to elevated ACTH. The patient was administered the intravenous isotonic infusion and the steroid treatment against an adrenal crisis. When the patient was evaluated with regard to adrenal insufficiency etiologies, the thyroid function and thyroid antibodies of the patient were negative, and she did not have type 1 diabetes, and parathyroid disease. Autoimmune adrenal insufficiency was not considered for the patient since there were not any accompanying autoimmune diseases. The patient, who had hilar enlargement in the lungs, were requester further examinations for tuberculosis. PPD was found anergic, the quantiferon test was negative, and the three consecutive says sputum ARB and TBC PCR results were negative. Tuberculosis was not considered primarily. A contrast-enhanced thoracic-abdominal computerized tomography scan was done with regard to other etiological reasons. In the CT scan, nodular consolidations, the largest being 27x27mm, in the bilateral lung upper lobes; lymph nodes, some containing calcifications and largest being 25x13 mm, in the bilateral hilar; a mass lesion, not presenting 39 HU density involvement before the contrast matter, 39 HU in the portal phase, and 38 HU in the late phase, containing millimetric calcifications with the size 29x26 mm in the right adrenal gland; a similar mass lesion with the size 26x15 mm at the left adrenal gland, not presenting 47 HU density washout before the contrast matter, and 57 HU density in the portal phase and the last phase were observed. PET-CT was requested considering malignity since the patient had a lesion in the lungs and massive lymphadenopathies.

In the PET-CT scan, pathologically enlarged FDG involvements were observed in the soft tissue in the velum with an approximate 17x10 mm size (SUV max: 6.4); in the bilateral submandibular and jugulodigastric lymph nodes (SUV max: 3.3), in the mediastinal region, the bilateral hilar and the lymph nodes (SUV max: 5.4), in the left superior lobe of the lung

(SUV max: 6.9), in the left adrenal gland (SUV max: 6.9), and near the left inferior quadrant bowel loops (SUV max: 9.5). PET-CT scan findings are shown in figure 1. In the colonoscopic examination intended for the involvement in the bowel loops, an irregular, fragile, rigid, and partly nodular view was observed along a 10 cm segment of mucosa in the transverse colon distal and the splenic flexura localization, and the biopsy results returned concordant with noncaseified epitheloid granuloma. A biopsy was made to the lesion with involvement in the velum, the result returned concordant noncaseified epitheloid granuloma. Sarcoidosis was considered primarily since the serum ACE of the patient was high (56.3 u/l), and the velum and colon biopsy results were concordant with noncaseified epitheloid granuloma.

Sarcoidosis involvement related primary adrenal insufficiency was considered since the patient did not have other etiological reasons for adrenal insufficiency. Biopsies were not taken from adrenal glands since it would be invasive and due to complication risks. The steroid dose started for the adrenal crisis treatment was decreased gradually, and the maintenance dose was continued. A dramatic amelioration in the symptoms of the patient was observed in the follow-ups.

Discussion

Sarcoidosis involvement in the endocrine organs is rarely observed. Thyroid, pituitary gland, and hypothalamus are most frequently involved among the endocrine organs.[3] Adrenal gland involvement, among endocrine organs, is very rarely observed. In cases with sarcoidosis adrenal involvement and when both adrenal glands are involved, generally adrenal insufficiency develops depending on the destruction of the glands. In single sided involvements, adrenal insufficiency is not expected, as in the Yu Sun Cha et al. case.

The adrenal insufficiencies observed with sarcoidosis in the literature are generally accompanied by polyglandular syndrome.[4, 5] In this case, autoimmune adrenal insufficiency was not considered primarily since the thyroid, parathyroid, and the pituitary functions

were normal, and she did not have type 1 diabetes and accompanying autoimmune diseases.

In the case of Kentaro Takahashi et al., another case not presenting autoimmunity in the literature, the buccal and gastrocnemius biopsy returned concordant with noncaseified granuloma of a patient presenting adrenal insufficiency and who had pathological involvement in the PET-CT scan, and the patient was diagnosed with sarcoidosis.[6] In our case, velum and rectum biopsies returned concordant with noncaseified granuloma. An adrenal malignity was not considered since the adrenal masses did not exhibit was out in the dynamic computerized tomography of the patient. We thought that the adrenal insufficiency setting could be related to sarcoidosis related involvement of the adrenal gland, after eliminating all other secondary reasons.

Conclusion

Although autoimmunity and tuberculosis are observed to a great extent in the etiology of adrenal insufficiency, it should be kept in mind that adrenal insufficiency can be related to sarcoidosis.

Declaration

The authors have no conflicts of interest to declare.

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