



OLGU SUNUMU/CASE REPORT

Spontaneous idiopathic bilateral adrenal hemorrhage in a pediatric patient

Çocukluk çağında idiyopatik spontan bilateral adrenal kanama

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Abstract

Adrenal hemorrhage occurs most frequently in the newborn, but it has been also well documented in older children and adults. There are several reasons in etiology and clinical symptoms can vary widely depending on the residual adrenal function. In this case; despite bilateral massive adrenal hemorrhage, no etiologic factor had been found and adrenal insufficiency was not present. To our knowledge, our patient is a spontaneous idiopathic bilateral massive adrenal hemorrhage case without adrenal insufficiency which is the first one reported in a pediatric patient in literature.

Key words: Spontaneous idiopathic bilateral adrenal hemorrhage, children, adrenal insufficiency

Öz

Adrenal kanama çoğunlukla yenidoğan döneminde görülse de daha büyük çocuklarda ve erişkinlerde de olabilmektedir. Etiyolojide birçok neden rol oynamakta ve klinik bulgu ve belirtiler çeşitlilik gösterebilmektedir. Sunulan vakada masif adrenal kanamaya rağmen adrena yetersizlik gözlenmemiş ve herhangi bir etiyolojik faktör bulunamamıştır. Bilgilerimize göre literatürdeki çocukluk çağında spontan idiyopatik bilateral masif adrenal kanaması olan ancak adrenal yetersizlik gelişmeyen ilk vaka olma özelliğini taşımaktadır.

Anahtar kelimeler: Spontan idiyopatik bilateral adrenal kanama, çocukluk çağı, adrenal yetmezlik

INTRODUCTION

Hemorrhage of the adrenal gland was first described by Canton in 1863¹. Early diagnosis is difficult because the clinical presentation may be nonspecific. Most cases are discovered at time of surgery or at postmortem examination and incidence in autopsy studies has ranged between 0.14% and 25%². It occurs most frequently in the newborn, but has been also well documented in older children and adults³.

It may result from trauma, acute stress, anticoagulation, coagulopathy, neonatal stress, underlying tumor, or idiopathic disease. Because its presentation is usually nonspecific and can easily be

mistaken for sepsis in a clinically ill patient; the diagnosis requires a high index of suspicion to prevent mortality.

In this case a patient with bilateral massive adrenal hemorrhage with neither adrenal insufficiency nor identified etiology is presented. It is the first one reported in a pediatric patient in literature.

CASE

A 14-year-old female presented with abdominal pain for two days and referred to our pediatric oncology department due to bilateral adrenal masses. On admission, physical examination was normal and her vital signs were also in normal ranges. In laboratory tests, only positive finding was low hemoglobin (8.9

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gr/dl) which was consistent with acute hemorrhage. In abdominal MRI bilateral large hematomas were seen 5.5x7 cm and 8,5x10 cm (Figure 1&2). There was no known history of recent trauma or chronic hematologic or other systemic disease. Patient was hospitalised. Erythrocyte transfusion was done only once when Hb value decreased down to 6.9 gr/dl. She was evaluated for surgery but conservative follow-up was found suitable due to high risk of morbidity of surgical exploration. Angiography was performed and there was no active bleeding, so embolization was not necessary (Figure 3). During etiologic research for infectious etiologies; blood and urine cultures, screening tests for TORCH, Hepatitis B, Hepatitis C, HIV, EBV, varicella, parvovirus infections, echinococcus granulosis (cyst hydatid) were done and resulted negatively. Tumor markers such as β -HCG, AFP, NSE, catecholamines (in 24-hour urine) were negative. Endocrinologic tests for adrenal insufficiency such as ACTH, cortisol, glucose and electrolytes were normal. To find out hematologic problems which could cause adrenal hemorrhage; coagulation parameters, d-dimer, homosistein, antiphospholipid and anticardiolipin antibodies, were obtained and all were negative. MTHFR 1298 gene mutation test result was homozygous normal. Malignancies such as neuroendocrine tumors and hematologic diseases were excluded. She was discharged finally. Her follow-up continued for two years and no problems had occurred.

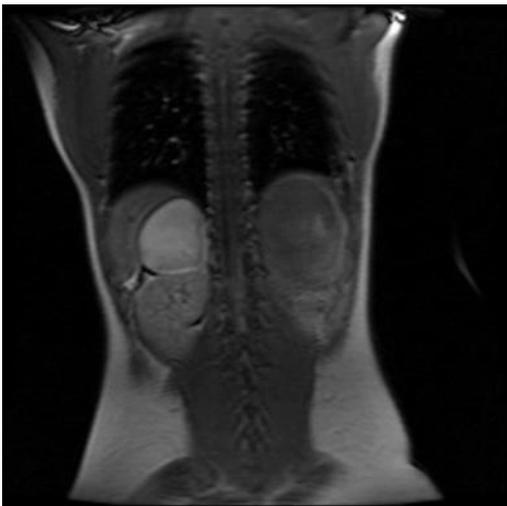


Figure 1- Coronal image from abdominal MRI of patient.

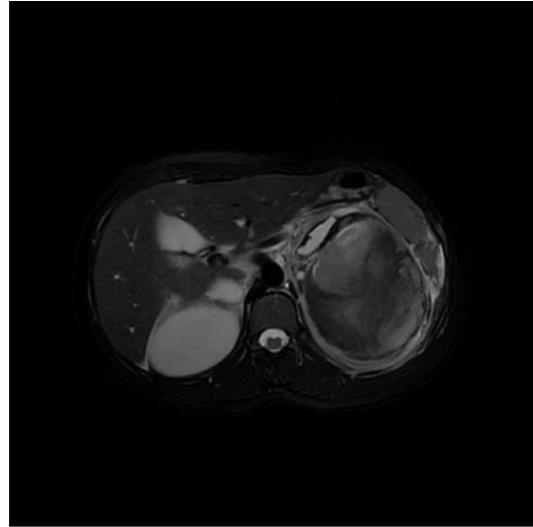


Figure 2. Axial image from abdominal MRI of patient.



Figure 3. Angiographic image of patient.

DISCUSSION

Adrenal hemorrhage in pediatric age group, occurs most frequently in the neonatal period and it is mostly associated with large fetal size, birth trauma owing to difficult labor especially breech presentation, perinatal asphyxia, septicemia or coagulation defects⁴. In older patients, there are several reasons in etiology such as trauma, acute

stress, sepsis (meningococcal is classical), anticoagulation, coagulopathy, prothrombotic states such as primary antiphospholipid syndrome (APS) and underlying tumors⁵. In our patient there was no history of trauma. Blood and urine cultures which were taken for several times were negative. Fever was thought to be due to adrenal hemorrhage itself since pyrexia is known to be a reliable sign of adrenal haemorrhage^{3,6}. Coagulation parameters were all normal and serologic tests for APS were negative. Adrenal tumors were excluded by USG, CT and MRI at presentation and during follow-up (only USG). Tumor markers were also negative. All these etiologies were excluded, making this case 'spontaneous idiopathic bilateral adrenal hemorrhage' which is the first one reported in a pediatric patient in literature.

The clinical presentation of adrenal hemorrhage varies from asymptomatic minimal bleeding, to fulminant hemorrhage with resultant death from hemorrhagic shock or adrenal insufficiency, depending on the rate of hemorrhage and residual adrenal function^{3,4,5}. Adrenal insufficiency signs become clinically evident if 90% of the adrenal cortex has been destroyed^{5,7}. Since most cases of adrenal hemorrhage are unilateral, a normal functioning contralateral adrenal gland prevents adrenal insufficiency. Patients may present with acute onset of abdominal pain, flank pain, nausea, vomiting, agitation, low-grade fever, dehydration, mental status changes, unexplained jaundice (newborn). In physical examination a palpable flank mass, hypotension, hypertension, scrotal hematoma (newborn) may be seen or it may be even totally normal. In a clinically ill patient, most of these symptoms could easily be mistaken for sepsis. Laboratory tests may be all normal or may show a significant drop in hemoglobin, hyponatremia, hyperkalemia, hypoglycemia and ketosis that could occur in adrenal insufficiency. Since adrenal insufficiency is a life-threatening clinical condition, timely diagnosis and treatment is essential⁵. In our patient both at presentation and during follow-up biochemical tests and blood gas parameters were in normal ranges. Endocrinologic tests were all normal, so it was presumed that adrenal insufficiency was not accompanying, despite bilateral massive hemorrhage.

In our patient surgical exploration was not found suitable and angiography was performed to see if there was active bleeding. There was no sign of

active bleeding and the decision was to continue conservative follow-up. It is widely agreed that surgical intervention for adrenal hemorrhage is not required if the patient is clinically stable^{5,8}. Nonoperative management includes supportive care, measurement of serial hematocrits and administration of blood transfusions if needed. Adrenal glands have complex vascular supply and embolization of one of the vessels usually does not lead to infarction. So embolization is a desirable treatment modality and alternative to surgical exploration in case of active bleeding^{5,9}.

Spontaneous idiopathic bilateral adrenal hemorrhage is an uncommon disease¹⁰ and there are no reports in pediatric population. Early diagnosis is difficult because the clinical presentation is nonspecific. Most cases are discovered only at time of surgery or at postmortem examination, so the diagnosis of this entity requires a high index of suspicion in a high-risk patient.

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