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Case Report

A Teenage Girl with Nonfunctioning Adrenocortical Carcinoma- A Rare Case

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Abstract

Adrenocortical carcinomas (ACs) are uncommon malignancies that can have protean clinical manifestations. A majority of cases are metastatic at the time of diagnosis, with the most common sites of spread being the local periadrenal tissue, lymph nodes, lungs, liver, and bone. Detection of tumors at an early clinical stage is crucial for curative resection. The present case was a 15 years old girl presented with abdominal distension and discomfort at Department of Surgery of Shaheed Ziaur Rahman Medical College and Hospital, Bogra, Bangladesh. The data were collected by history taking, clinical examination, laboratory investigations, trans-abdominal ultrasonographic examination, Computed Tomography of abdomen and by histopathological study of the excised surgical specimen. From histopathological report the case was finally diagnosed as adrenocortical carcinoma. This case report emphasize the significance of thorough evaluation of all women presented with vague abdominal pain. Although this condition is rare, it is potentially dangerous in its massive form and hormonal effect as the prevalence is more in female than male..

Key words: *Nonfunctioning adrenocortical carcinoma, Teenage girl, CT scan*

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Introduction

Adrenocortical carcinomas are rare aggressive tumors. Their annual incidence is approximately one to two per million among the population of the United States of America.[1] Approximately 60% of adreno-cortical carcinomas are functional tumors.[2] The female-to-male ratio for ACs is approximately 2.5-3:1. The accumulation of data, especially in international registries, revealed the incidence of adrenal tumors to be higher in female individuals than had previously been thought, particularly in those aged 0-3 years and those over 13 years. Nonfunctional ACs are distributed equally between the sexes. AC occurs in 2 major peaks: in the first decade of life and again in the fourth to fifth decades. While, functional tumors are more common in children, however, nonfunctional tumors are more common in adults. Based on data from the International Pediatric Adrenocortical Tumor Registry, the median age at which children develop adrenal carcinomas is 3.2 years; 60% are younger than four years, and 14% are older than 13 years.[2-4] Patients with active endocrine tumors often present with cushing's syndromes alone (45%), a mixed cushing's and commonly present with an enlarging abdominal mass and abdominal or back pain or with an incidental finding on radiographic imaging called an "adrenal incidentaloma". Laboratory investigations include the following serum glucose, serum cortisol, serum adrenal androgen, urine adrenal hormone, urine vanillylmandelic acid (VMA) and urine homovanillic acid (HVA) levels. Imaging studies include Xray abdomen in erect posture, ultrasonography, Computed tomography (CT) scanning and magnetic resonance imaging (MRI) but CT, MRI are the imaging studies of choice in AC. The typical imaging feature is characterized by mass with irregular edges in adrenal region. The presence of contiguous adenopathy serves as corroborating evidence. Some of the macroscopic features of an AC that suggest malignancy include a weight of more than 500 g, the presence of areas of calcification or necrosis, and a grossly lobulated appearance.[5-8] Histological findings also include numerous mitoses, scant cytoplasm, and none of the rosettes observed in neuroblastoma. When feasible, total resection remains the management modality of choice for the definitive treatment of AC. It also remains the only potentially curative therapeutic modality. While open laparo-tomy for adrenalectomy represents the standard of care, several reports suggest a role for laparoscopic resection if the adrenal tumor is small and there is no preoperative evidence of metastatic disease. Medical care in patients with AC, which can be supportive or adjuvant to surgical resection,

encompasses treatment of endocrine excess syndromes, use of mitotane or several multiagent chemotherapy regimens, treatment with prevention of potential complications, strategies for palliative and terminal care issues, including symptom relief and management.[1, 3, 9-11] A variety of staging system has been used for ACs. The Union of International Cancer Control (UICC) proposed the first TNM classification of malignant tumors for AC in 2003. However, an analysis based on data from the German AC Registry revealed several shortcomings of this classification system. Therefore, ENSAT developed a revised staging system. The superiority of the ENSAT staging system over the 2004 UICC/American Joint Committee on Cancer Classification System for prognostication was confirmed in a recent North American study. Estimated five-year disease-specific survival rates of patients with stage-1 and stag-4 cancer in the studies were 82% and 13%, respectively.[9,10] Nowadays, adreno-cortical cancer is often diagnosed after a great delay, when the cancer is very advanced. The only potentially curative treatment for ACC is surgical resection,[11] which is technically possible in most patients with stage-1 and 2 diseases. The most important predictor of survival in patients with adrenal cancer is adequacy of resection. Patients who undergo complete resection have five years actuarial survival rates ranging from 32% to 48%, whereas median survival is less than one-year in patients who undergo incomplete excision. Other treatment options include treatment with mitotane, an adrenocorticolytic drug, as well as adjuvant chemotherapy and palliative irradiation.

Case Report

A 15 years old girl with a history of abdominal pain and swelling in epigastric, left hypochondriac, and left lumbar region was admitted in surgery ward at Shaheed Ziaur Rahman Medical College Hospital, Bogra, Bangladesh. The swelling presents for last four months with occasional abdominal pain which was dull aching, non-radiating and aggravates by taking food. She had early satiety and nausea for same duration. The patient had no previous medical diseases or surgical operations. Her menarche commenced at the age of 12-years with irregular cycle, duration and heavy flow. But she had no menstruation for the last two years. She denied uses of any medications. Her bladder and bowel habit were normal. General examination revealed, the patient was anxious looking, below average nutrition, anaemic, blood pressure 90/60 mm of Hg with all other parameters were normal. Her weight was 35 kg, height was 140 cm and the secondary sex characteristics were well developed. On abdominal examination, left flank was full

with a lump which was globular in shape, smooth regular surface, hard in consistency, non-tender, moved with respiration and not fixed with overlying skin. Pelvic and digital rectal examination revealed normal. She had no lymphadenopathy or hepatosplenomegaly and no clinical signs of hormonal excess. The patient was advised for all relevant laboratory investigations, hormonal evaluations, ultrasonography of whole abdomen and CT scan of abdomen. She had raised ESR (60 mm in 1st hour), Hb-7.0 g/dl and hyponatremia (Na-130 mmol/L). Others laboratory findings with hormonal evaluation revealed normal. Ultrasonography of whole abdomen revealed a retroperitoneal mass inseparable from left kidney and spleen was not identified (Figure 1).

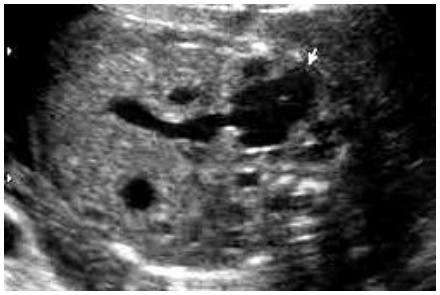

Figure 1: Ultrasonography revealed heterogeneous mass with central necrosis in left adrenal region

CT scan of abdomen showed a retroperitoneal mass originated from upper pole of left kidney pushing the left kidney downwards, spleen laterally, stomach medially and as suggested mass arising from left adrenal gland with differential diagnosis of enlarged lymph node (Figure 2).

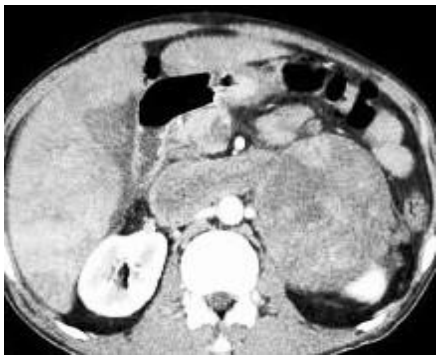

Figure 2: **CT scan revealed heterogeneous enhancing mass lesion with central necrosis in left adrenal region pushing the left kidney downwards**

During operation, left subcostal incision was given and a mass which measured about 12×15 cm (Figure 3) was removed. The mass was sent for histopathological examination, the features of that mass were consistent with adrenocortical carcinoma. After operation the patient was managed postoperatively with all subsequent measures.

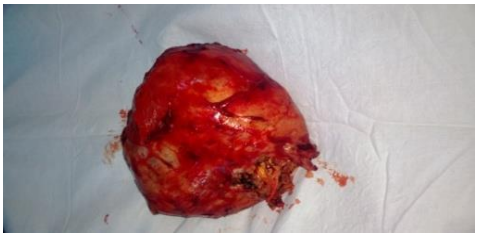

Figure 3: Tumor after resection

Discussion

Approximately 60% of adrenocortical carcinomas are functional tumors.[2] Patients with active endocrine tumors often present with Cushing's syndromes alone (45%), a mixed Cushing's and virilization syndrome

(25%), or virilization alone (<10%).[3] Conversely, patients with nonfunctioning tumors more commonly present with an enlarging abdominal mass and abdominal or back pain. Our present case presented as nonfunctional tumour with abdominal pain which was dull aching, non-radiating and aggravates by taking food, early satiety and pain and swelling in epigastric, left hypochondriac, and left lumber region. Patient had no lymphadenopathy or hepato-spleenomegally and no clinical signs of hormonal access. The accumulation of data,[2-6] especially in international registries, revealed the incidence of adrenal tumors to be higher in female individuals than had previously been thought, particularly in those aged 0-3 years and those over 13 years. Present case was 15 years old girl and age-gender demography was comparable with previous studies. A hormonal work-up for functional ACs is widely mandatory; however, the question whether to perform this evaluation in apparently asymptomatic patient has been debated.[4] Plasma metanephrine level or urinary metanephrine and catecholamine levels may be measured to exclude pheochromocytoma. Plasma aldosterone and rennin levels may be measured in patients with hypertension and/or hypokalaemia. In present case laboratory investigations were within normal limit. In this study ultrasonography revealed a retro-peritoneal mass inseparable from left kidney but spleen was not identified. CT scan of abdomen showed a retro-peritoneal mass originated from upper pole of left kidney pushing the left kidney downwards, spleen laterally, stomach medially and as suggested mass arising from left adrenal gland with differential diagnosis of enlarged lymph node. Previous studies[3,4,7] revealed CT scanning can detect adrenocortical mass and also distinguish adenomas from ACs.[3,4,7] The size of adrenal mass visualized on imaging studies was the single most important criterion to help diagnose malignancy. In the series reported by Copeland PM,[5] 92% of adrenal tumors were greater than 6 cm in diameter. MRI was complementary to CT in that local invasion and involvement of the vena cava were more readily identifiable. A fine-needle aspiration biopsy cannot distinguish a benign adrenal mass from a metastatic tumor. Capsular or vascular invasion was the most reliable sign of cancer. In the absence of these findings, the Weiss histopathological system is the most commonly used method for assessing the likelihood of malignancy because of simplicity and reliability.[6,7] Immunohis-tochemistry is also helpful in rendering the diagnosis.[5, 8]

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