

Adrenocortical Carcinoma – A Case Report

Dr. (Mrs.) M. M. Paithankar* Dr. Deepti Deshmukh** Dr. Rashmi Nagdeve** Dr. Neetu N. Agrawal#

* Associate Professor, Department of Medicine Government Medical College, Nagpur

** Lecturer, Department of Medicine Government Medical College, Nagpur

Post graduate student, Department of Medicine Government Medical College, Nagpur

ABSTRACT:

We report a case of an 18 year old female who presented with paraparesis as the initial manifestation of adrenocortical carcinoma. This case report being reported for rarity of the adrenocortical carcinoma and its unusual manifestation in the form of paraparesis.

KEY WORDS: Adrenocortical carcinoma, paraparesis

Introduction:

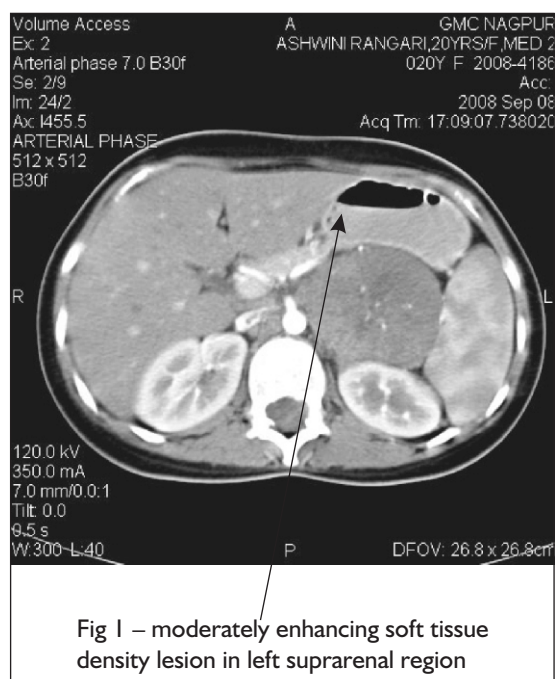
Adrenocortical carcinoma (ACC) affects one or two out of every million people per year. There is a bimodal age distribution, with peaks in the first and fourth decades. In adults, the mean age at diagnosis is approximately 45 years. Most series report a slight female predominance. A significant number of patients (30–85%) have distant metastases at presentation, and the majority of these patients will not survive much beyond 1 year. The majority of patients with adrenocortical carcinoma presents with advanced disease and have an extremely poor 5-year survival rate.¹ We hereby report a case of adrenocortical carcinoma who presented unusually for the first time as paraparesis.

Case Report:

An 18 year old female, presented with fever since 25 days associated with backache. She noticed weakness in both lower limbs while walking since 15 days. There was no history of sensory complaints or bowel and bladder involvement. There was history of anorexia and weight loss. There was no history of cough/night sweats. Patient denied any history of headache or abdominal pain. There was no history of tuberculosis in past. Her family history was not significant. On examination she was febrile (100.7°F). Her pulse was 110/min and BP was 120/70 mm of Hg. She had pallor but no icterus, cyanosis or clubbing. There was no lymphadenopathy. She had spinal tenderness at around D₇ & D₈ level. On per abdominal examination, there was no organomegaly or lump. No abnormality was detected on respiratory and cardiovascular examination. She was conscious, oriented and there was no cranial nerve abnormality. Hypertonia was present in both lower limbs and power was grade 4. Upper limbs examination was normal. Deep tendon reflexes in lower limbs were exaggerated and both

plantars were extensor.

On investigations, her hemoglobin was 9.4 gm%, peripheral smear showed normocytic and hypochromic red cells, adequate platelets and no hemoparasite. Her kidney and liver function tests were within normal limits. No organisms were grown on blood culture. To our surprise, her USG abdomen showed retroperitoneal mass lesion 7.6 X 9.7 X 5.5 cm in left side of upper abdomen. CT Abdomen showed moderately enhancing soft tissue lesion in left suprarenal region with multiple lytic lesion involving T8 vertebral body with associated paravertebral and intraspinal soft tissue extension with significant canal compromise – possibility of Neuroblastoma or Paraganglioma (Figure 1 & 2). Her urinary VMA levels were in normal range (2 mg in 24 hrs).



CT guided FNAC from the mass was done which was suggestive of pleomorphic population of round to

nephrectomy. Patient is at present on radiotherapy for spinal metastasis.

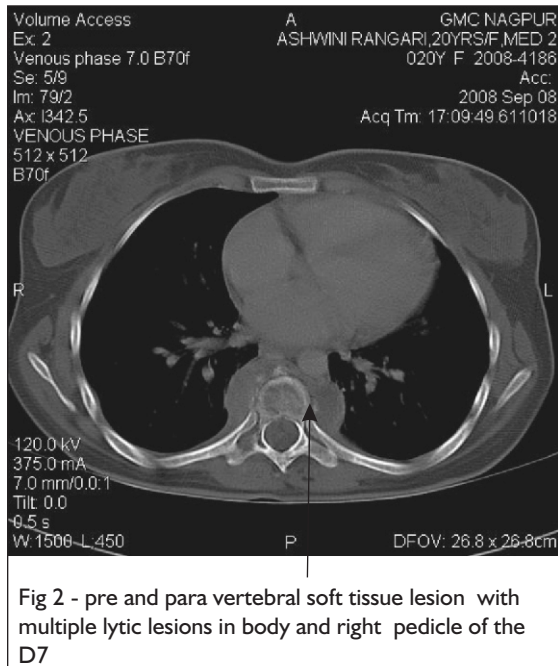


Fig 2 - pre and para vertebral soft tissue lesion with multiple lytic lesions in body and right pedicle of the D7

polygonal cells in clusters & dispersed singly. Cells had variable amount of cytoplasm with well defined borders and nuclei were pleomorphic with abnormal chromatic pattern, large nucleoli & mitotic figures s/o adrenocortical carcinoma. Patient was then taken up for surgery and mass was removed along with splenectomy and left sided

Discussion:

The hormonally active variants of ACC constitute approximately 60% of cases while rest 40% remains hormonally silent. This second variety typically present with fever, weight loss, abdominal pain and tenderness, back pain, abdominal fullness, or symptoms related to metastases or the mass is found incidentally, during either examination or radiologic imaging.

ACC, generally, carries a poor prognosis and is unlike most tumours of the adrenal cortex, which are benign (adenomas) and only occasionally cause Cushing's syndrome. Five-year disease-free survival for a complete resection of a stage I-III ACC is approximately 30%. The most important prognostic factors are age of the patient and stage of the tumor. The best chance of survival was achieved by surgical extirpation with the value of adjuvant therapy yet to be determined.²

Oida T, et al reported a case of adrenocortical carcinoma who presented with fever and malaise as the initial manifestation.³ Presentation of adrenocortical carcinoma as paraplegia is yet unreported to the best of our knowledge. The purpose of reporting this case is to highlight rare presentation of a very rare malignancy.

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