



Case Report

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A Rare Case of Adrenocortical Carcinoma Recurrent and Disseminated

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Abstract

Adrenocortical Carcinoma is an extremely rare tumor and a few cases so far been reported in the literature. We hereby present one case report. A 38-Year-old Indian female house wife presented with a complaint of occasional Headache and left flank pain last two to three months. Associated with weakness, fever, and anorexia. She had history of left Adrenalectomy on 15.08.2018. Her routine investigation and CT Thorax and abdomen MRI lumbosacral spine, USG abdomen and pelvis, immune histopathology and tumor marker were performed, diagnosis of recurrent Adrenocortical carcinoma with Lung, liver spleen and retroperitoneal lymph nodes metastasis was established. Staged IV according to AJCC. Performance score ECOG -2. We planned palliative chemotherapy with best supportive care. She was discharge in stable condition with follow for next cycle.

Keywords: Adrenocortical carcinoma; Cachexia; Polymenorrhoea; Cortisol; Immunohistochemistry; Adrenalectomy

Abbreviations: ACC; CT; DHEAS; ACTH; ECOG; AJCC; LN; ENSAT

Introduction

Adrenocortical Carcinoma is a very rare disease. The incidence is 1 to 2 per million per year [1]. The median is of diagnosis is fifth to sixth decade of life [2]. The ratio of female and male ranges from 1.5:1. Aside from genetic (Li Fraumeni syndrome, Lynch Syndrome) predisposition no risk factors have been established. According to national modality follow back Survey (1986) smoking is Male and contraceptive in Female before age of 25 is also risk factors [3]. Patient many presented with complain of the clinical features of hormone excess and present with nonspecific symptoms like local tumor growth, abdominal pain, abdominal fullness, early satiety [4] and rarely presented with cachexia and night sweats, Paraneoplastic syndromes [5]. Hormone relative symptoms also appear when there excess in hormone-secreting ACCs. At the time of diagnosis, the initial evaluation should include a thorough physical examination, detail history. Basic biochemical evaluation includes RFT, LFT, CBC, Hormone assay. Imaging modalities include CT, MRI of abdomen/pelvis and CT Thorax.

For staging of tumor we followed AJCC, TNM Staging :-

Stage 1:- Tumor 5cm or less with no spread to surrounding tissues or lymph nodes without distant metastasis.

Stage 2:- Tumor >5 cm with other stage 1 characteristics.

Stage 3:- Tumor invading nearby tissues and/or spread to nearby lymph nodes.

Stage 4:- Distant metastasis.

In our case study in stage 4

There is newly introduced European Network for the study of Adrenal Tumor (ENSAT) system become widely used by ACC community. Stage group as Table 1

Table 1: Staging System for ACC.

	AJCC	ENSAT
Stage-1	T1 N0 M0	T1 N0 M0
Stage-2	T2 N0 M0	T2 N0 M0
Stage-3	T1-2 N0 M0 , T3N0M0	T1-2 N1 M0 , T3-4N0M0
Stage-4	T1-4 N0-1 M1 , T3-4 N1 M0 , T4 N0 M0	T1-4 N0-1 M1

She was diagnosed in 2016 outside the hospital August 2016. Patient was evaluated with a CECT Scan which showed enhancing mass lesion in Lt Suprarenal with multiple areas of Necrosis.

Left adrenal not separately visualized. Inferiorly abutting upper left kidney, no infiltration, Sub cm LN seen in retroperitoneum along the celiac artery. Calcific segment 7 of Liver. ? Adrenocortical carcinoma. Patient underwent left Adrenalectomy on 15.08.2018, histopathology report shown gross-capsulated soft tissue mass 500 gm 11x9x6.8 cm. Histology shows tumor cells round to polygonal with arrangements of sheets, Extensive necrosis and tumor emboli seen in sinusoids and a thin-walled vessel in capsule with Capsular invasion, Mitosis 3-4/50 HPF. Histological feature in favor of Adrenocortical carcinoma 5 Celiac lymph nodes with reactive Hyperplasia. Pan CK, Vimentin, chromogranin and CD10 are negative. Carelentin, Melan A are positive. KI 67+10-15% post of USG was normal, no residual mass.

Post OP DHEA

52.3. Patient has been on follow-up with serial USG which is insignificant, follow-up USG done on 11.06.2019 was s/o an ill-defined mass lesion 4.6*5 cm left suprarenal region ? Recurrence Patient also complains of Polymenorrhoea. Cervical pep smear done in December 2018 which is negative for malignancy. USG done December 2018 showed thickened endometrium with small follicles in ovaries b/L.? Anovulatory cycles.

Examination

ECOG-1, Patient well oriented to time, place, person, pulse rate 86/minute, BP-180/110 mm Hg., RR-18/minute., P/A-Soft, Vague mass palpable in left suprarenal region, no orgenomegaley, Prominent abdominal aortic pulsations present, Post OP Scar Health. No dehiscence.

Investigation

CBC, RFT, LFT, Urine VMA, Serum, Cortisol, Serum ACTH, Se. Electrolytes.

CT Thorax and abdomen

Multiple parenchymal nodules and masses of varying sizes are seen in both lungs, largest measuring 3.5x3.4x3.0 cm (APxTRxCC) in lower lobe of left lung. There is a large lobulated heterogeneously enhancing mass seen in left suprarenal region. Left adrenal is not seen separately. It shows loss of fat planes with left crus of diaphragm medially. Posteriorly it is involving posterior pre renal space and infiltrating posterior renal fascia. Upper pole of left kidney is not demarcated separately likely involve the mass. Surgical staples are seen engulfed within the lesion. Anteriorly there is focal loss of planes with spleen. Surgical staples also noted in retroperitoneum. A small deposit is seen in right perihepatic region along segment VI. Minimal free fluid in POD with mild enhancement and modality? Metastasis

MRI Lumbosacral Spine

Variable disk desiccation small well defined suspicious non enhancing T1/T2/STIR hypointense signals noted in L4 vertebral body, no evidence of involvement of posterior and

lateral elements. Lung, liver spleen and retroperitoneal lymph nodes metastasis.

Management

As she presented to us with metastatic disease (Liver, Lung, Spleen, Retroperitoneum Lymph nodes Metastatic). We thoroughly evaluate the patient by physical, clinical examination and essential biomarker with imaging and start palliative chemotherapy (Etoposide, Carboplatin, Doxorubicin with G-CSF support). After two chemotherapy we further evaluate for the patient for response. In evaluation diseases progressive in nature and we planned for tablet metotan which is not available in market hence we continue the palliative chemotherapy with the best supportive management.

Conclusion

Adrenocortical Carcinoma is a very rare neoplasm. More common in female. ACC with disseminated metastases has very poor prognosis. Hence early diagnosis and complete tumor resection is only curative intent. Adjuvant chemotherapy helps improve survival and decrease the chance of recurrence and relapse rates. Multiple disseminated metastasis has no significant advantage of chemotherapy but a recent phase 3 trial (FIRM- ACT, First International Randomized Trial in Metastatic Adrenocortical carcinoma Treatment) EDPM regimen is most efficient. Single agent Mitotane can be considered as palliative intent. Over all the ACC is a rare neoplasm and require early detection, correct diagnosis and multimodality approach including total excision, adjuvant chemotherapy.

Discussion

Adrenocortical carcinoma is a rare malignancy with worst prognosis with incidence is 1 to 2 per million per year. More common in female (58.7%Predilection) [1], fifth to sixth decade of life. ACC occurs with increased frequency in Beckwith Wiedman syndrome and Li-Fraumani syndrome. However , most of the tumors are sporadic [6]. Diagnosis will be established by clinical, biochemical markers, imaging and histological features. ACC has unfavorable prognosis. Tumor characteristics of malignancy and velocity of tumor growth are usually related to a decreased survival. Distant metastasis and number of organs involved in metastatic disease, confers a poor prognosis [7]. Only curative approach to ACC is complete tumors resection. The evidence that Surgery for recurrent disease indicated in when disease confined to one site or organ [8]. Adjuvant Chemotherapy aim to minimize the chance of recurrence. In unrespectable or metastatic ACC palliative care and palliative chemotherapy should be consider. Mitotane remain the only drug approved by U.S. Food and Administration for adjuvant and palliative indent [9]. Dose of mitotan 1g. twice daily with increase every 4 to 7 days by 0.5 g/d till maximum 5 to 7 g. [10] Chemotherapy is currently a mainstay of treatment for advanced and metastatic ACC. EDPM (Etoposide, doxorubicine, Cisplatin, Mitotan) regimen is most effective regimen [11].

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