A Case Report on Adrenocortical Carcinoma

Preksha Aghara, Shaini Shah*, Minesh Gandhi**, Cherry Shah****

Abstract

Adrenocortical carcinoma (ACC) is a rare malignant neoplasm arising from adrenocortical parenchymal cells with an aggressive biological behavior. A 60 years old male patient came to general surgery OPD of tertiary care hospital with chief complaint of sudden weight loss in a month associated with mild abdominal pain. On abdominal sonography approximately 10x8x8 cm sized mixed echogenic lesion with internal vascularity was noted in right suprarenal region. Open right adrenalectomy was performed. Based on overall histopathology,it was concluded that adrenocortical carcinoma of right adrenal gland. The Weiss criteria of adrenocortical malignancy found as the most reliable tool for histopathological scoring system. Most ACC cases are hormonally functional, and immunohistochemical analysis of steroidogenic enzymes has provided pivotal information as to the analysis of intratumoral production of corticosteroids.

Key Words: Adrenocortical carcinoma, Immunohistochemistry, Steroidogenesis, Weiss criteria.

Introduction:

Adrenocortical carcinoma is rare malignant epithelial tumor of adrenal cortical cells, with an annual incidence of 0.7 to 2 per million.[1] The Weiss criteria of adrenocortical malignancy found as the most reliable tool for histopathological scoring system.[2] ACCs are more common in females than in males. The tumor has bimodal age distribution with one peak in children of less than 5 years of age and one peak in adults in fourth or fifth decade.

Case Report:

A 60 years old male patient came to general surgery OPD of tertiary care hospital with chief complaint of sudden weight loss in a month associated with mild abdominal pain. Not associated with burning Micturition, fever, nausea, vomiting, etc. Hormonal serum cortisol level was 6.12 microgram/dl suggested that hormonally tumor was inactive. X-ray showed radio-opacity in right hemi pelvis. On abdominal sonography approximately 10x8x8 cm sized mixed echogenic lesion with internal vascularity was noted in right suprarenal region. Open right adrenalectomy was performed. The mass was sent to histopathology department. Grossly, specimen was received in multiple fragmented yellowish soft tissue portions measuring 15x12x2.5 cm, weighing around 300 grams. The largest tissue portion was measuring 13.5x12x2.5 cm.On its cut surface yellowish whitish area with variegated appearance were seen. Microscopic examination revealed trabecular and solid sheets of tumor cells. Tumor cells showed nuclear pleomorphism, hyperchromasia, atypical 8-10 mitotic figures/50 HPF and bizarre tumor cells. There were presence of marked areas of confluent necrosis, hyaline globules, degenerative changes and tumor giant cells. Infiltration into the capsule and surrounding adipose tissue with vascular invasion were present. Based on overall histopathology applying modified weiss criteria it might be concluded that ADRENOCORTICAL CARCINOMA OF RIGHT ADRENAL GLAND,PATHOLOGICAL STAGE : T3NxMx, stage-III.

Immunohistochemistry shows positivity for Ki67 index and vimentin.

* Resident
** Associate Professor
*** Professor & Head, Department of Pathology, Smt. NHL Municipal Medical College, Ahmedabad, Gujarat, India
Correspondence : Dr. Preksha Aghara
E-mail : preksha.aghara1510.ap@gmail.com

Figure 1: 15×12×2.5 cm variegated multiple yellowish soft tissue portions

Figure 2: Atypical mitotic figures along with vesicular & pleomorphic nuclei

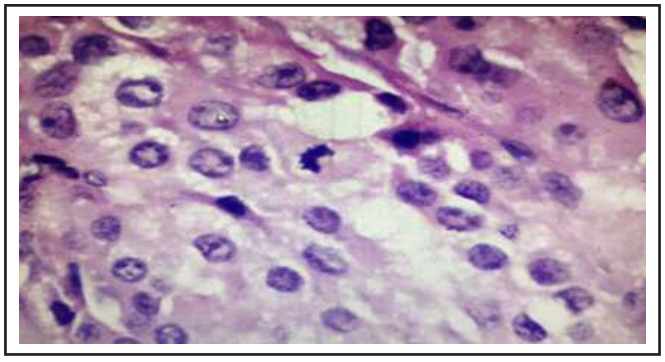


Figure 3: Capsular & vascular invasion with tumor cells

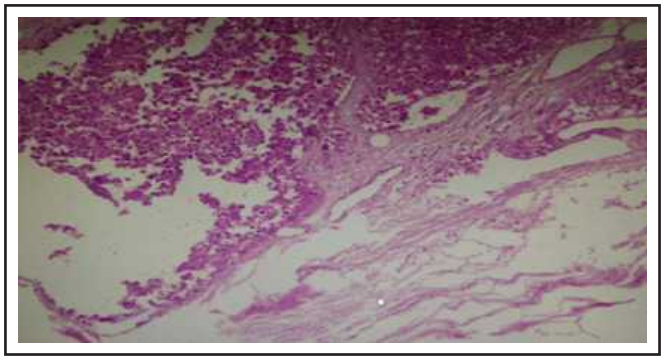


Figure 4: Degenerative necrotic areas with hyaline globules

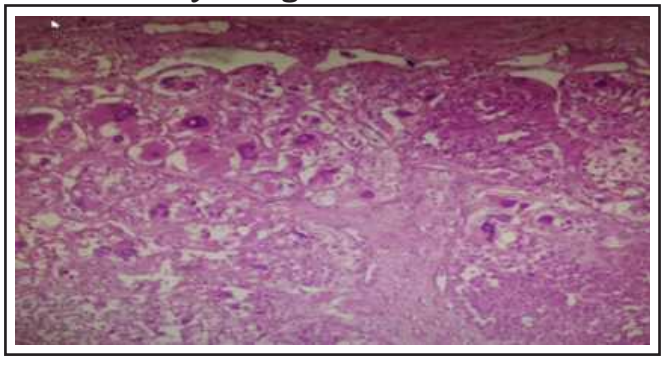


Figure 5: Ki-67 index positive

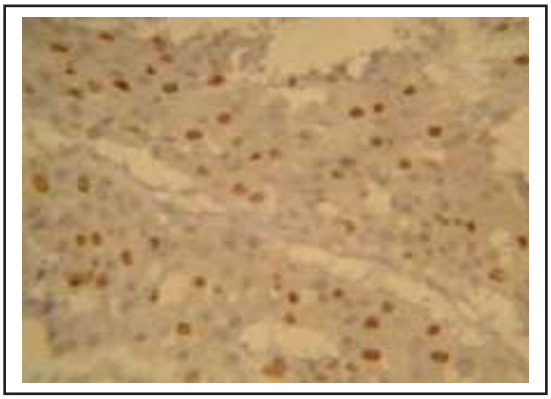
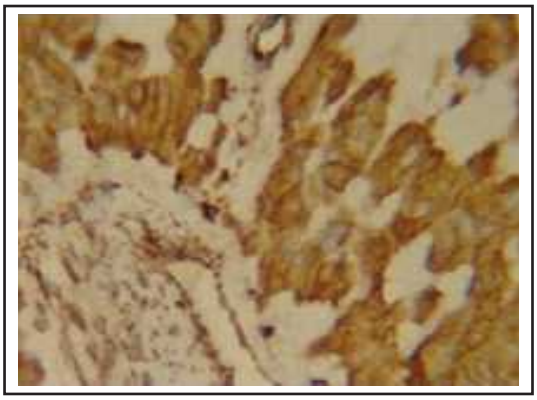


Figure 6: Vimentin positive



Discussion:

The evaluation and categorization of adrenocortical neoplasms remain among the most challenging areas in adrenal pathology. 50% are functional and associated with virilizing and other syndromes but it cannot determine function from morphology.[3] Hormonally inactive ACCs usually present with gastrointestinal symptoms or back pain caused by mass effect.[4] High level of DHEA-S is suggestive of adrenocortical carcinoma.

The differential diagnosis of ACC includes pheochromocytoma, adrenal adenoma and renal cell carcinoma. In pheochromocytoma tumor cells arranged in cell nests – Zell ballen pattern with positivity for chromogranin & synaptophysin. In adrenal adenoma tumor cell with bland, round nuclei and positive for calretinin & inhibin.[4] Renal cell carcinoma contains glands which may contain RBCs, glycogen & keratin positive.

Only definitive criteria for malignancy are distant metastasis or local invasion. The Weiss scoring system, first introduced in 1984, has been widely used for the histologic diagnosis of ACC.[5] The nine histological parameters evaluated in this system are high nuclear grades, mitotic rate >5/50 high power fields, atypical mitotic figures, clear tumor cell cytoplasm, diffuse architecture, necrosis, venous invasion, sinusoidal invasion and capsular invasion.[5] A tumor is labeled malignant when it meets 4 or more of these histological criteria.Modified Weiss criteria includes mitotic rate >5/50 hpf, clear cytoplasm in tumor cells, atypical mitosis, necrosis and capsule invasion. The tumor, lymph node & metastasis (TNM) classification system proposed by the AJCC is the most frequently used classification to assess the local extension of the primary tumors, lymph nodes involvement and the presence of distant metastasis. The TNM classification is one of the most important prognostic tools.

Mete et al. emphasized that among all criteria angio invasion, characterized by tumor cells invading through vessel wall and /or intravascular tumor cells admired with thrombus is the most important biological parameter in ACCs.

Immunohistochemical analysis using a comprehensive panel of markers including vimentin, synaptophysin, Ki-67, Melan-A, calretin, CK and EMA may be required for the confirmation of diagnosis of ACC.[6]

Conclusion:

On applying gross and microscopic findings into the Weiss criteria, any adrenal tumor can be diagnosed as benign or malignant which can be confirmed by immunohistochemistry. Our case report establishes the importance of the Weiss system and tumor size, weight as an important parameters in predicting malignant behavior of ACC.

References:

1. Erickson LA, Rivera M, Zhang J. Adrenocortical carcinoma: review and update. AdvAnatPathol 2014 May; 21: 151-9.
2. Aubert S, Wacrenier A, Leroy X, et al. Weiss system revisited: a clinicopathologic and immunohistochemicalstudy of 49 adrenocortical tumors. Am J SurgPathol 2002; 26: 1612-29.
3. Erickson LA. Challenges in surgical pathology of adrenocortical tumors. Histopathology 2018 Jan; 72: 82-96.
4. Duan K, Giordano TJ, Mete O. Adrenal cortical proliferations. In: Mete O, AsaSL, eds. Endocrine pathology. UK: Cambridge University Press, 2016; 602-27.
5. Weiss LM. Comparative histologic study of 43 metastasizing and nonmetastasizing adrenocortical tumors. Am J SurgPathol 1984; 8: 163-9.
6. Mete O, Asa SL, Giordano TJ, et al. Immunohistochemical biomarkers of adrenal cortical neoplasms. EndocrPathol 2018 Jun; 29: 137-49.