Rhabdoid adrenocortical carcinoma with brain metastasis: A case report

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ABSTRACT

Introduction: Adrenocortical carcinoma (ACC) is a rare disease with the incidence about 0.02-0.2% from all case of malignancies. The number of malignancies cases reported annually being about 0.7-2 cases per million people. ACC can be functional or nonfunctional tumors, being diagnosed incidentally because of loco-regional symptoms or distant metastases. The overall 5-year survival rate after diagnosis was 15% to 47% and there are no significant differences in survival based on patient age, gender or tumor functional status.

Case Report: A 47 years old male came into emergency room with nausea, vomiting, headache and weakness for 4 weeks. From the physical examination, there were slight hypertension and tenderness on left hypochondric region. Abdominal ultrasound showed a solid mass with central necrotic on upper left abdominal cavity. Then, contrast abdominal CT revealed a well defined heterogeneous solid mass, contrast enhancement with necrotic area that attached on the upper pole of left kidney suspicious on left adrenal organ. He was performed left radical adrenalectomy as initial treatment to determine the histopathology result and tumor staging. After surgery complains of his headache remained, thus head CT was performed and it concluded an intracranial metastatic process at right parietal region, left temporal brain parenchymal and cerebellum which pressing ventricle IV. Steroid and neurotrophic treatment were given by neurologic division. This patient was also consulted to neurology, hemato-oncology, neurosurgery and radiotherapy for further treatment.

Conclusion: ACC is a progressive disease with a poor prognosis because of its initial presentation with advance disease (tumor burden or distant metastases), high incidence of local recurrence, and systemic therapy still controversial. The patient has to do a complete physical examination, imaging and laboratory work-up. Multimodality treatment with a multidisciplinary approach is needed to manage the patient for optimal outcome.

Keywords: Adrenocortical carcinoma, adrenal tumor, adrenalectomy, pheochromocytoma.


INTRODUCTION

Adrenocortical Carcinoma (ACC) is a rare disease with the incidence about 0.02-0.2% from all case of malignancies. The number of malignancies cases reported annually being about 0.7-2 cases per million people. ACC can involve both children and adults, with median age about 56 years old (between 1-91 years old). Commonly, ACC was found incidentally because of loco-regional symptoms or distant metastases. Differential diagnosis between primary ACC, such as adrenocortical adenoma, pheochromocytoma, renal cell carcinoma and metastatic tumor is hard to be done, determination is based on precise correlation between clinicopathological and immunohistochemical result.4,4

Lymph node metastases are very rare to be found, but the systemic symptoms are experienced by half number of patients.4 The prognosis of this tumor is poor, because it is an advanced disease with only 15% to 47% of overall 5-year survival rate and no significant differences in survival based on gender, patient age or tumor functional status.4,6

In this report, we present a case of Rhabdoid ACC in a 47 years old man which found incidentally from gastritis symptoms and brain metastases from headache without complete laboratory test. Radical adrenalectomy combined with multimodality treatment was done as standard plan of care.

CASE REPORT

A 47 years old male came into emergency room with nausea, vomiting, palpitation, headache and weakness for 4 weeks. From the physical examination, there were a slight hypertension and tenderness on left hypochondric region, left temporal brain parenchymal and cerebellum which pressing ventricle IV. Steroid and neurotrophic treatment were given. Endoscopy and abdominal ultrasound had been done. Erosive esophagitis, bile reflux and superficial antrum gastritis were found by endoscopy. Abdominal ultrasound concluded a solid mass,
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9.3 x 8.0 cm, with central necrotic process at upper left of abdominal cavity, well defined and flat edge which hard to differentiate with the border of upper pole left kidney; intratumoral vascularisation expressed in colour doppler ultrasound (CDUS). Thus, contrast abdominal CT was performed and revealed a well-defined heterogeneous solid mass, 99.2 x 101.8 x 112.1 mm, flat edge with necrotic area that attached on superior of left renal and heterogeneous contrast enhancement. (Figure 1).

Open left radical adrenalectomy with retroperi-toneal approach was performed as initial treatment to determine the histopathology result and tumor staging. Macroscopic examination of the surgical specimen showed most of the tumor was necrotic and bleeding. It contained many necrotic tissues, hoop of tumor had cracked and along the tumor there was a frail tissue mass. Small mass tumor appearance, brownly colour, irregular with bleeding focal, specimen showed 13x10x6 cm in size. Microscopic examination from tumor mass showed malignant epithelial cells, epithelial cells with sinesisia cytoplasm, hyperchromatic core, prominent nucleoli vesicular, bleeding between malignant cells, polyhedral eosinophil cytoplasmic and a group of malignant cells which looked rhabdoid appearance (Figure 2A-D).

After operation, patient still complained of his headache so head CT was performed and it concluded an intracranial metastatic process at right parietal region, left temporal brain parenchyma and cerebellum which pressing ventricle IV (Figure 3).

Multidiscipline approaches have been prepared for this patient, such as consulting to neurology, hemato-oncology, neurosurgery, radiotherapy department and also palliative treatment.

DISCUSSION

Adrenocortical tumors commonly located at adrenal cortex. It can be functional or nonfunctional tumor. The use of work-up imaging has been enhancing the incidental detection of ACC, despite, the majority of patients start complaining when they are in an advanced stage of disease. In this report, patient showed more nonfunctional symptoms such as nausea, vomiting and headache. The tumor was also found incidentally through abdominal ultrasound and he already in advance stage which the tumor already metastasizes into the brain.

Adrenocortical carcinomas are usually big with irregular borders on CT. Size is vital in diagnosing these tumors. By the time they are found, they are often 12 cm in diameter. Over 90% of adrenocortical carcinomas are >6 cm at presentation. Our cases, we found the tumor was large, 10x10x11 cm. Beldegrun et al. also stated that 92 % of mass which greater than 6 cm were adrenal carcinomas. But in other series, David et al. stated that 16% of the carcinomas could be <5 cm in diameter.

Tumor staging is a widely used tool to assess prognosis in patients with cancer. The size of patient tumor was 13 cm which obtained after operation and there was a distant metastasis into the brain, so based on UICC (Union Internationale Contre Le Cancer) staging system, patient is at stage IV of ACC with <5% of 5 year survival rate. Whereas
stage I and stage II are defined as a localized tumors with size \( \leq 5 \) and \( >5 \) cm. Stage III ACC is featured by infiltration of surrounding adipose tissue, tumor thrombus in the vena cava or positive regional lymph nodes.\(^{12,13,15}\) The overall survival for ACC patient is poor, 45-60\% when ACC is detected in early-stage (stage I and II) and 10-25 \% in advanced stage (stage III and IV).\(^{15}\)

The most treatment approach for ACC is complete tumor resection followed by adjuvant therapies. These two approaches used mainly to treat ACC.\(^{9,12}\) Euddeum shim et al. also stated that the best treatment approach is complete surgical resection but there is no establishment of adjuvant chemotherapy.\(^7\) According to Hemsen et al., there is a high chance of survival when doing complete surgical resection. After complete resection, the median survival number reaches 27.6-74 months and 7.6-15.9 after incomplete resection.\(^8\) In our cases, tumor resection was done in this patient by doing open radical adrenalectomy. Operation was successfully done without any hemodynamic complication. In metastatic case of ACC, if there is one nodule or single lobe of metastasis in brain or lung, can be treated by metastasectomy.\(^5\)

Then, patient was given corticosteroid therapy by neurologist and was planned to achieve adjuvant chemotherapy and radiotherapy for brain metastases. The rate of local recurrence ranges between 19 to 34\% even though complete surgical resection has been done, because of that, adjuvant therapy can be given as a solution for recurrent tumor or the tumor is unresectable. One of adjuvant chemotherapy which is widely used is mitotane.\(^{12}\) Radiotherapy also significantly lowered the risk of local recurrence progression in patients with ACC but there is no evidence in improving the overall survival.\(^{9,15}\)

After discharged from hospital, patient is scheduled for follow up, but he already died 4 months after operation before received any chemotherapy and radiotherapy. Mauricio et al. reported 6 patient ACC with brain metastases in his clinical series, whose three of them died, two patients is alive and one was lost to follow up which patients had received chemotherapy, underwent metastasectomy and radiosurgery.\(^{11}\) Different result was also reported in Hemsen et al. case series. Six patients with recurrent and metastatic ACC to liver and lung were treated with surgical resection and adjuvant therapy which all of them still alive. But the prevalence of these patients with long survival is low.\(^9\)

Treatment of ACC often includes multimodal therapy directed by team of surgeons, medical oncologists, endocrinologists and radiotherapy oncologists.\(^7,15\) As ACC is an aggressive malignant tumor, patients should be followed every 3 months during and after initial treatment.\(^{12}\) Detection of tumor in early clinical stage is mandatory in all case of ACC so complete resection can be done earlier for better survival rate outcomes.\(^{14}\)

**CONCLUSION**

ACC is a progressive disease with a poor prognosis because of its initial presentation with advance disease (tumor burden or distant metastases), high incidence of local recurrence, and systemic therapy still controversial. The majority of patient usually come to doctor already in high stage of tumor or the tumor found incidentally. The patient should undergo complete physical examination, imaging and laboratory work-up. Multimodality treatment and multidisciplinary approach are needed to manage the patient for optimal outcome. Any further research is also required to be done.

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**CONFLICT OF INTEREST**

The author declares there is no conflict of interest regarding publication of current case report.

**ETHICAL CONSIDERATION**

The patient has received signed informed consent regarding publication of their medical data in journal article before any data collection.

**REFERENCE**


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