



Oncology

Adult renal neuroblastoma: A case report

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ARTICLE INFO

Keywords:

Cancer
Kidney
Neuroblastoma
Adult
Nephrectomy
Chemotherapy

ABSTRACT

Adult renal neuroblastoma is a rare condition, and a few cases have been previously described in the literature. Its prognosis is variable and the treatment suffer the lack of specific guidelines due to the low incidence. We report a case of adult renal neuroblastoma managed with nephrectomy and adjuvant chemotherapy. The purpose of this report is to update the knowledge available on this disease.

1. Introduction

Neuroblastoma is a cancer arising from cells of the neural crest that form the adrenal medulla and sympathetic ganglia. This cancer may occur anywhere along the sympathetic chain and mainly affects the adrenal glands and retroperitoneum.¹ Neuroblastoma is typically a childhood cancer. More than 90 % of cases of the tumors occur in children under 10 years old.² Renal neuroblastoma is uncommon, particularly in adults. In 2018, Huang conducted a review of all published cases highlighting the diagnosis, management and outcome over time.³ We report a case of adult renal neuroblastoma treated with nephrectomy and adjuvant chemotherapy.

2. Case presentation

A 26-year-old male consulted for a left lumbar mass evolving for 2 months. No significant medical history was found. There was no fever, hematuria, diarrhea or vomiting and the blood pressure was normal. The physical examination revealed a left upper quadrant mass, extended to the epigastrium and the periumbilical region, sensitive, with firm consistency and irregular contours. Urinalysis was negative. Laboratory tests showed microcytic hypochromic anemia (hemoglobin 9.28 g/dL) and a normal kidney function. Abdominal ultrasonography showed a heterogeneous mass in the upper pole of the left kidney. CT scan showed

a large hypodense mass without calcification on non-enhanced images, measuring 10.4 x 20.9 x 21.1 cm, in the upper pole of the left kidney. Enhanced images showed heterogeneous enhancement and centrally necrotic mass. Lumboaortic, mesenteric and inguinal lymph nodes were noted. There was no evidence of metastatic disease.

The patient underwent a left nephrectomy with lymph node dissection. During exploration, there was a large left kidney tumor that pushed back the digestive organs (Fig. 1). The adrenal gland was normal and preserved. The removed tumor weighed 500 g and measured 37 x 26 x 11 cm (Fig. 2). The postoperative period was uneventful and the patient was discharged on postoperative day 5. Pathological examination revealed that the tumor was compatible with differentiated neuroblastoma (Fig. 3) with an intermediate Mitotic Karyorrhectic Index (2–4%). According to the International Neuroblastoma Staging System (INSS), the patient was classified as stage 3 at initial presentation. After surgery, the patient received 3 courses of chemotherapy with doxorubicin and carboplatin. The 6-month follow-up CT scan showed metastatic recurrence (liver, lung and pelvic lymph nodes) and the patient died one month later.

3. Discussion

Adult renal neuroblastoma is a rare condition. The first published case was presented by Baumgartner in 1975(4). A review of case reports

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<https://doi.org/10.1016/j.eucr.2023.102614>

Received 3 November 2023; Accepted 8 November 2023

Available online 15 November 2023

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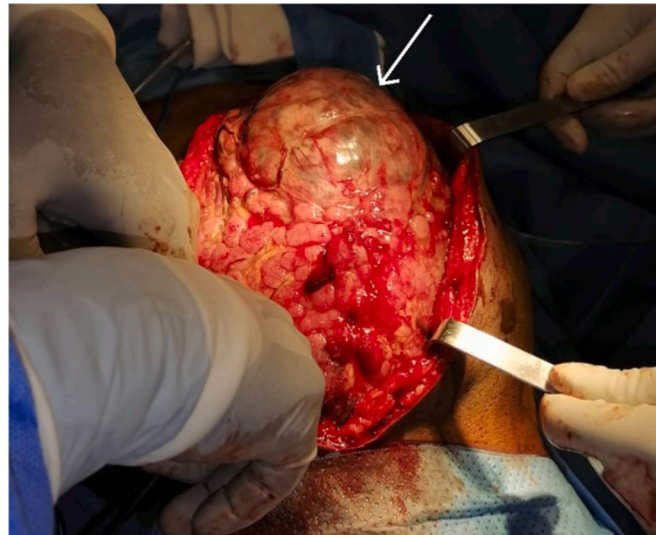


Fig. 1. Intraoperative exploration: tumor of the upper pole of the left kidney (arrow).



Fig. 2. Removed renal mass.

in the literature was presented by Huang in 2018. He reported 8 cases published worldwide including his own case.³ The clinical manifestations of renal neuroblastoma vary widely. The symptoms include: hematuria, renal mass, abdominal mass, pain, and hypertension as part of the paraneoplastic syndrome.^{1,3} In our case, the patient presented with lumbar mass as described in the literature.⁵⁻⁷

CT scan is the gold standard for the evaluation of renal tumors (size, location, relationship to the upper urinary tract and vessels).⁸ According to the current literature,³ the CT findings include: isodense renal mass without calcification on nonenhanced CT; heterogeneously enhancing

mass with central necrosis after contrast medium injection, associated with perirenal invasion, regional lymphadenopathy, or renal vein thrombosis. In our case, the CT findings were similar to those in the literature, which can lead to misdiagnosis with a renal cell carcinoma. However, other tests may help differentiate neuroblastoma from renal cell carcinoma prior to surgery. Those modalities include: fine-needle aspiration cytology, catecholamine metabolites tests, and meta-iodobenzylguanidine (MIBG) scintigraphy.^{1,3}

Due to the lack of specific guidelines, the treatment of adult renal neuroblastoma is based mostly on the recommendations for neuroblastoma in children. According to all published cases,^{3-7,9,10} treatment modalities are surgery alone, or surgery plus adjuvant therapy (chemotherapy or radiotherapy). Huang and colleagues presented the case of a 41-year-old female with stage 4 neuroblastoma.³ She was treated with surgery and 6 courses of adjuvant chemotherapy with cisplatin, doxorubicin, etoposide, and cyclophosphamide. There was no evidence of recurrence during a 24-month follow-up period. In our case, the patient was treated with surgery and 3 courses of adjuvant chemotherapy with doxorubicin and carboplatin. However, like the second case of Gohji, chemotherapy was not effective and the patient's condition gradually worsened.⁵ The 6-month follow-up CT scan showed metastatic recurrence and the patient died one month later. The prognosis was poor due to delayed diagnosis at advanced stage and poor performance status which affects response to chemotherapy.

4. Conclusion

Adult renal neuroblastoma is uncommon. Due to this low incidence, the diagnosis and treatment of adult renal neuroblastoma are complicated. Further study is required to provide specific guidelines for better management of this disease.

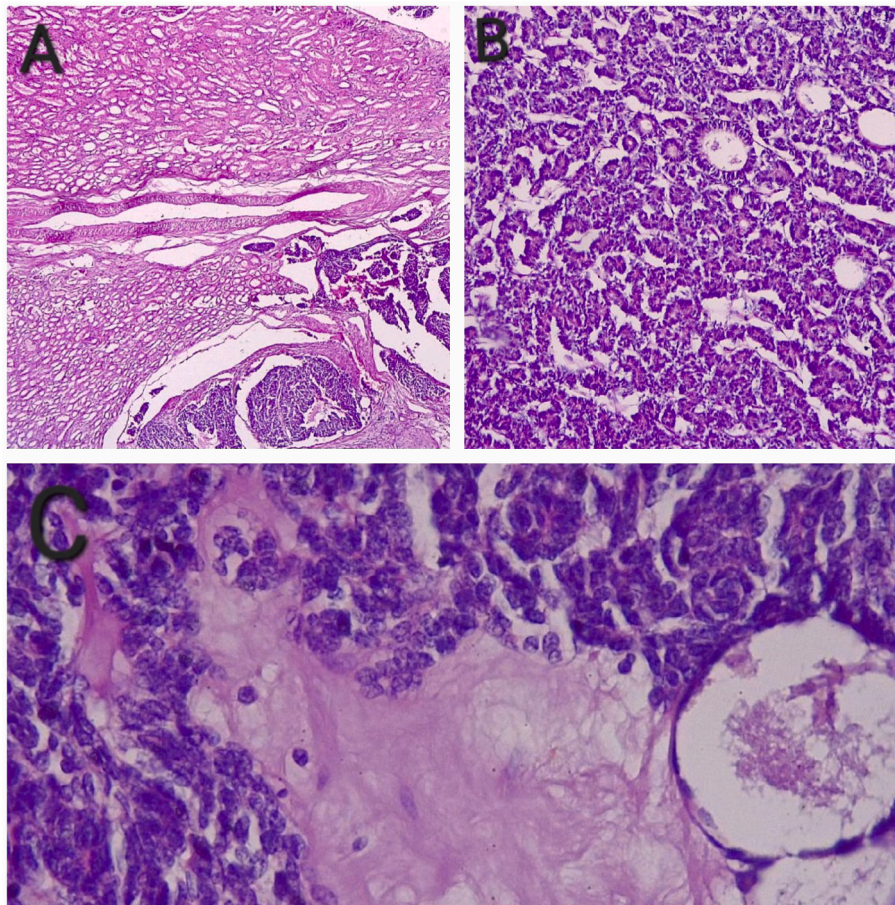


Fig. 3. Pathological examination (hematoxylin and eosin staining) showed: (A) renal tumor with small cells, granular chromatic nuclei and scanty cytoplasm (x200) ; (B) the cells are arranged as rosettes (x250) ; (C) neuropil presence (x300)..

Ethical approval

The Hospital Ethical Committee gave the agreement to report this case.

Funding

The authors declare they have received no funding for the preparation of this document.

Author contribution

These authors participated in the making and correction of this document. All authors agreed with the publication of the document.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Declaration of competing interest

The authors report no declarations of interest.

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