

Adult intrarenal neuroblastoma presenting as renal cell carcinoma

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Introduction

Neuroblastoma is known to arise from cells of the neural crest that form the adrenal medulla and sympathetic ganglia. Tumours may occur anywhere along the sympathetic chain within neck, thorax, retroperitoneum, pelvis or in the adrenal gland. Seventy-five percent arise in the retroperitoneum, 50% in the adrenal, and 25% in the paravertebral ganglia.¹ The variety of locations in which these tumours can arise and the spectrum of their differentiation result in a wide range of clinical presentations and behaviours.¹ These tumours can undergo spontaneous regression, differentiate to benign neoplasm² or exhibit extremely malignant behaviour.³

Neuroblastoma has a variable prognosis and is very rarely reported in adults. There are only 3 cases in the English literature on kidney neuroblastoma. We present a case of neuroblastoma in a woman who had an intrarenal mass.

Case report

A 37-year-old female was referred to our clinic by a nephrologist for evaluation of renal mass. Abdominal ultrasonography showed a 81 × 61 mm mass in the right kidney. Her physical examination revealed right upper quadrant mass. She denied significant loss of weight. Urinalysis demonstrated microscopic hematuria and leucosuria. Laboratory evaluations revealed hemoglobin 12.4 mg/dL, and her creatinine level was 0.61 mg/dL. Computerized tomography (CT) showed a 70 × 60 mm regular, heterogenous and centrally necrotic mass in the lower pole of the right kidney (Fig. 1, Fig. 2). Both chest radiographs and bone scan showed no evidence of metastatic disease.

The patient had undergone right nephroureterectomy and ipsilateral adrenalectomy with anterior subcostal incision.

There was no adhesion to the adjacent areas and the tumour was removed intact and completely. On gross examination the tumour was 60 × 60 × 60 mm and located in the lower pole. Macroscopically, the tumour was surrounded by the renal capsule and was divided clearly from the normal renal tissue of lower pole. Cut surface of the tumour was yellowish, and necrotic tissue was seen at the centre of the tumour. Microscopically, the tumour was composed of small cells with hyperchromatic nuclei and scanty cytoplasm, arranged as rosettes. For the differential diagnosis, such as primitive neuroectodermal tumours and Wilms' tumour, several immunohistochemistry staining were performed. Tumour cells were reacted with synaptophysin, whereas CD-99, chromogranin, WT-1, neuron specific enolase (NSE) and pancytokeratin were found negative (Fig. 3, Fig. 4, Fig. 5). These microscopic and immunohistochemical findings revealed that this tumour was compatible with neuroblastoma.

The postoperative period was uneventful and the patient was discharged on postoperative day 3. Bone marrow biopsy was performed on the 45th day after the operation and found normocellular bone marrow. The patient was assumed as stage 1 disease. Six months after the operation, there were no abnormal findings on her CT. The patient received no adjuvant therapy, and she had no evidence of recurrent residual disease 30 months after surgery.

Discussion

Neuroblastoma is a rare malignancy with about 500 cases diagnosed each year in the United States.⁴ The incidence is 2.26 per million person-years, with almost all cases diagnosed before the age of 10 years. The incidence rate for patients aged 30 to 39 is 0.2 cases per million person-years,⁴ but kidney neuroblastoma is very rarely reported in adults.

There are no treatment recommendations for neuroblastoma in adults. The treatment modalities in children are surgery, chemotherapy and radiation therapy. The role of each of these modalities varies depending on tumour stage,



Fig. 1. Axial computerized tomography image of the tumour.



Fig. 2. Axial computerized tomography image of the tumour.

age and biologic prognostic factors. These characteristics can be used to stratify patients into favourable and unfavourable categories by risk group. Children with stage I have a disease-free survival rate of greater than 90% with surgical excision alone.³ Adjuvant therapy (chemotherapy with or without radiotherapy) is reserved for recurrence or higher stage tumours with MYCN amplification and/or unfavourable histologic features.⁵

Primary neuroblastoma of the kidney had not been described in the literature until 1986. Kazuo Gohji and colleagues reported two unusual cases of neuroblastoma of the kidney in adults.⁶ In both patients right radical nephrectomy was performed. Microscopically, both tumours were composed predominantly of scant pink cytoplasm and round nuclei with rosette formation with interspersed fibrils. The first patient received 3000 rad of irradiation to the tumour

bed and para aortic area. There was no evidence of tumour recurrence at follow-up. Progressive disseminated disease was documented in the second patient despite postoperative adjuvant chemotherapy with combined cisplatin and epipodophyllotoxin, and combined vincristine, cyclophosphamide, doxorubicin and dimethyl-triazeno imidazole carboxamide. Chemotherapy was not effective and the patient's condition gradually worsened.⁶

McLean and colleagues presented the case of a 39-year-old man with stage 1 neuroblastoma with unfavourable histologic features; he was treated with surgery alone.⁷ No treatment recommendations existed for the treatment of neuroblastoma in adults, so they treated this patient according to pediatric guidelines. The patient received no adjuvant therapy, and he had no evidence of recurrent of residual disease 21 months after surgery.⁷ Like this case, we treated our patient with surgery alone. The patient received no

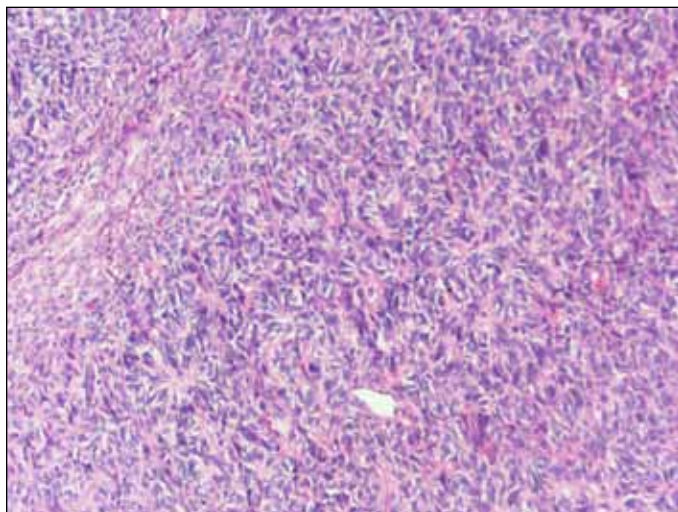


Fig. 3. This microphotograph shows tumour cells with hyperchromatic nuclei and scanty cytoplasm, are concentrically arranged around eosinophilic fibrillary materials (neuropil) (hematoxylin & eosin stain x 200).

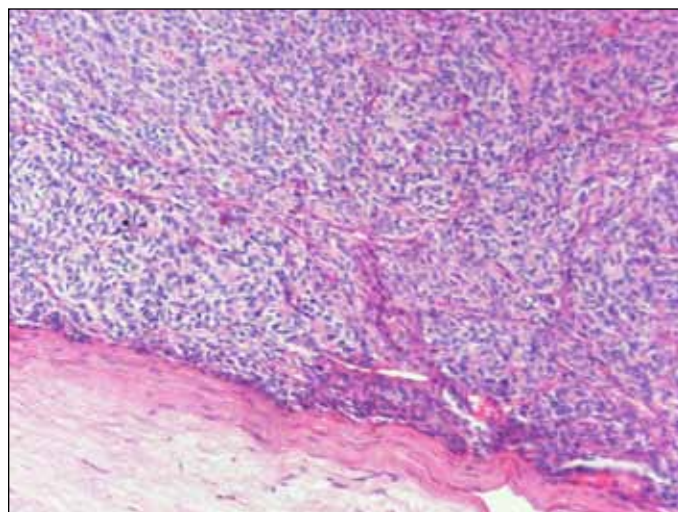


Fig. 4. This picture shows encapsulated circumscribed tumor (hematoxylin & eosin stain x 200).

Table 1. Main clinicopathological findings in previously reported cases and present case

Cases	Age	Laterality	Tumour size (cm)	Placement of tumour in the kidney	Treatment	Follow-up
Case 1, Gohji ⁴	35	Right	15×11×10	Lower pole	Surgery + radiotherapy	No recurrence
Case 2, Gohji ⁴	29	Right	28×25×20	Upper pole	Surgery + chemotherapy	Patient's condition had been gradually worsening
Case 3, McLean ⁵	39	Right	9	Upper pole	Surgery alone	No recurrence (21 month after surgery)
Case 4, Present case	37	Right	6×6×6	Lower pole	Surgery alone	No recurrence (30 month after surgery)

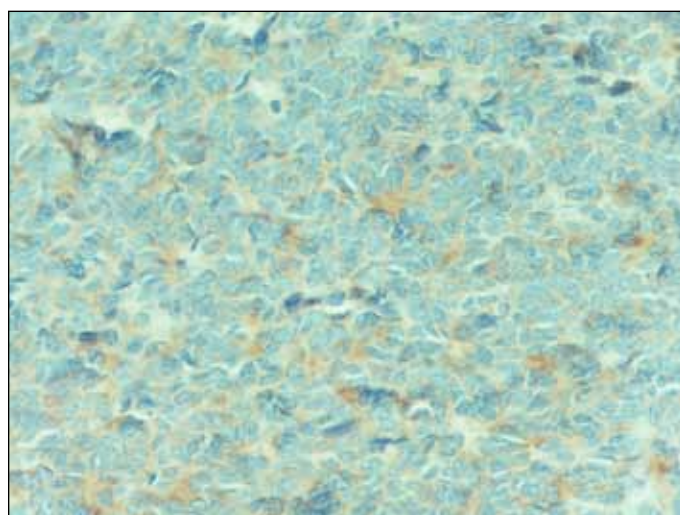


Fig. 5. Immunohistochemical staining revealed that tumour cells, especially neuropil in the centre of rosettes were reacted for synaptophysin (synaptophysin x 200).

adjuvant therapy, and she had no evidence of recurrent of residual disease 30 months after surgery. Main clinicopathological findings in previously reported cases and our present case are summarized in Table 1.

Conclusion

Neuroblastoma is extremely rare in adults. There are only 3 cases of kidney neuroblastoma in the literature. However,

in patients undergoing radical nephrectomy for renal mass, renal neuroblastoma should be included in the differential diagnosis. If these tumours are limited to the kidney, prognosis is favourable after radical nephrectomy.

Competing interests: None declared.

This paper has been peer-reviewed.

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