

Primary Renal Rhabdomyosarcoma in An Adult Patient Presenting With Myasthenia Gravis: Unusual Presentation of A Rare Malignancy



Medical Science

KEYWORDS : Rhabdomyosarcoma, renal cell carcinoma, myasthenia gravis, radical nephrectomy, immunohistochemistry

Prakasa R B

Department of Urology, Government General Hospital, Guntur Medical College, Guntur, Andhra Pradesh

Rambabu B

Department of Urology, Government General Hospital, Guntur Medical College, Guntur, Andhra Pradesh

Sridhar P

Department of Urology, Government General Hospital, Guntur Medical College, Guntur, Andhra Pradesh

ABSTRACT

Genitourinary rhabdomyosarcoma is commonly seen in children and is rare in kidneys. Renal sarcomas account for less than 1% of cases of renal cell carcinomas. Myasthenia gravis is rarely seen as a paraneoplastic syndrome in renal cell carcinomas. Renal rhabdomyosarcoma in adult is hence a rare entity with only three or four cases reported in the literature. Usually the diagnosis is made postoperatively after histopathologic examination and immunohistochemistry of the radical nephrectomy specimen. Here we present a case of primary renal rhabdomyosarcoma in an adult who presented with myasthenia gravis as a paraneoplastic syndrome. A radical nephrectomy was done with careful perioperative use of acetylcholine esterase inhibitors. Histopathologic features were suggestive of mesenchymal tumour with skeletal muscle differentiation and positive staining for desmin and myogenin indicating a rhabdomyosarcoma.

INTRODUCTION

Rhabdomyosarcoma is a tumour of mesenchymal origin with skeletal muscle differentiation and is common in children. Renal sarcomas account for only 1% of cases of renal malignancy. Primary renal rhabdomyosarcoma is very rare in adults. Here we present a case of renal rhabdomyosarcoma in an adult patient who presented with myasthenia gravis as a paraneoplastic syndrome and was treated by a radical nephrectomy.

CASE HISTORY

A 30 year old male presented with a one month history of abdominal distention and pain involving right side of the abdomen. There was no history of haematuria or fever. There was history of loss of weight but no loss of appetite. He had history of drooping of eyelids and gave a family history of ptosis suggesting a possibility of congenital myasthenia gravis. Neurologic evaluation revealed a decremental nerve response suggestive of neuromuscular junction disorder. Ach receptor antibodies were positive suggestive of myasthenia gravis. Per abdominal examination revealed an ill-defined mass involving right hypochondriac and right lumbar regions. The mass was bimanually palpable.

Ultrasonography revealed a heterogeneous mass lesion arising from right kidney. Contrast enhanced CT revealed a mass arising from right kidney with heterogeneous enhancement and cystic areas. There was no renal vascular invasion. There were no pulmonary or liver metastases (Figures 1, 2).

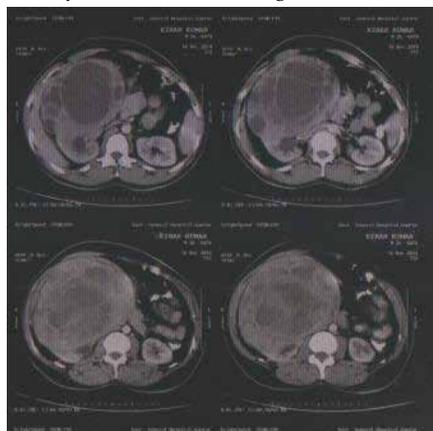


Figure 1: CECT images

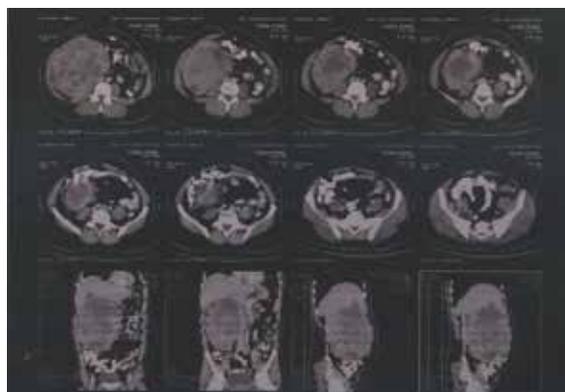


Figure 2: CECT images

Patient underwent a right radical nephrectomy under careful perioperative use of acetylcholine esterase inhibitors. There was a tumour arising from the lower pole of right kidney measuring 20x15 cm with variegated consistency. There was no renal vein invasion and no infiltration of surrounding structures. Hilar lymph nodes were present and a lymphadenectomy was done.



Figure 3: Intraoperative photo

On pathological examination cut surface showed haemorrhagic and cystic areas. Microscopically highly pleomorphic cells with eosinophilic cytoplasm and prominent nucleoli were found with multinucleated tumour giant cells and bizarre strap cells interspersed (Figures 4, 5). Lymph nodes showed reactive lymphadenitis. Immunohistochemistry for vimentin and myogenin was

positive suggesting a myogenic origin of the tumour.

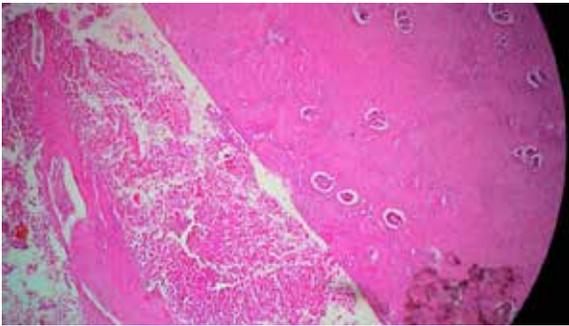


Figure4: Tumour with adjacent normal kidney

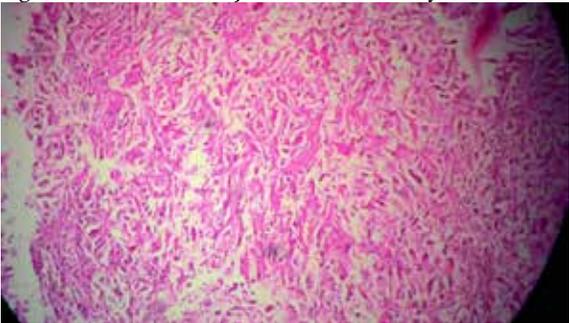


Figure 5 : pleomorphic spindle shaped cells and strap cells

DISCUSSION

Rhabdomyosarcomas are high grade small round blue cell tumours with skeletal muscle differentiation. They are among the more common tumours occurring in children but are rare in adults. Four variants have been described – alveolar, embryonal, pleomorphic and botryoid (1). Pleomorphic type is common in adults with predominant distribution in the limbs (2). Geni-

tourinary rhabdomyosarcomas are common in paratesticular region, other common regions being bladder, prostate and female genitourinary tract. Renal sarcomas are as such rare tumours accounting for 1% of all renal malignancies (4). Leiomyosarcoma is the most common sarcoma with cell of origin being smooth muscle cell of capsule or perinephric structures (5). Renal rhabdomyosarcomas are rare tumours of mesenchymal origin arising from renal parenchyma. Renal rhabdomyosarcomas have poorer prognosis than other genitourinary sites of rhabdomyosarcomas. Though there are several paraneoplastic syndromes in renal cell carcinoma, myasthenia gravis has not been reported as one previously in the literature. Hence this is a rare way of presentation of a rare malignancy.

Renal sarcomas are difficult to differentiate clinically and radiographically from renal cell carcinomas. Clinical presentation may include palpable mass, hematuria, abdominal pain which are non-specific. Radiographic features also may be confusing with renal cell carcinoma with large soft tissue mass replacing kidney which may show poor or heterogenous contrast enhancement (6). Usually the diagnosis is made postoperatively after histopathologic examination of the specimen. Immunohistochemistry with positive staining for Myogenin is a sensitive and specific marker for diagnosis of rhabdomyosarcoma indicating a skeletal muscle origin and differentiation of the tumour (7). Radical nephrectomy is the treatment of choice and the prognosis of rhabdomyosarcoma in adults appears to be poorer as compared to children.

CONCLUSION

Primary renal rhabdomyosarcoma is a rare histopathologic diagnosis of a renal cell malignancy. Myasthenia gravis may be a paraneoplastic syndrome in renal malignancy and careful perioperative management is required. There is insufficient data and literature at present to recommend the use of adjuvant chemotherapy or radiotherapy in these patients after radical nephrec-

REFERENCE

- [1] R. C. Horn Jr. and H. T. Enterline, "Rhabdomyosarcoma: a clinicopathological study and classification of 39 cases," *Cancer*, vol. 11, no. 1, pp. 181–199, 1958 | [2] D. Dalfior, A. Eccher, S. Gobbo et al., "Primary pleomorphic rhabdomyosarcoma of the kidney in an adult," *Annals of Diagnostic Pathology*, vol. 12, no. 4, pp. 301–303, 2008. | [3] Kaplan WE, Firlit CE, Berger RM; Genitourinary rhabdomyosarcoma : *J Urol*. 1983 Jul;130(1):116-9. | [4] V. Srinivas, P. C. Sogani, S. I. Hajdu, and W. F. Whitmore Jr., "Sarcomas of the kidney," *Journal of Urology*, vol. 132, no. 1, pp. 13–16, 1984 | [5] Deyrup AT, Montgomery E, Fisher C . Leiomyosarcoma of the kidney: a clinicopathologic study; *Am J Surg Pathol*. 2004 Feb; 28(2):178-82 | [6] N. Lalwani et al. "Pediatric and adult primary sarcomas of the kidney: a cross-sectional imaging review," *Acta Radiologica*, vol. 52, no. 4, pp. 448–457, 2011 | [7] M. H. Cessna et al., "Are Myogenin and MyoD1 expression specific for rhabdomyosarcoma? A study of 150 cases, with emphasis on spindle cell mimics," *American Journal of Surgical Pathology*, vol. 25, no. 9, pp. 1150–1157, 2001. |