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ISSN: 0976-3031

International Journal of Recent Scientific Research
Vol. 6, Issue, 6, pp.4407-4409, June, 2015

International Journal of Recent Scientific Research

RESEARCH ARTICLE

PRIMITIVE NEUROECTODERMAL TUMOUR OF KIDNEY WITH RIGHT ATRIAL THROMBOSIS

Mhaske S.M, Patwardhan S.K, Naveen T, Patil B and Gajengi A

Department of Urology Seth G.S. Medical College Mumbai, Maharashtra, India

ARTICLE INFO

Article History:

Received 14th, May, 2015
Received in revised form 23th, May, 2015
Accepted 13th, June, 2015
Published online 28th, June, 2015

Key words:

Kidney, Primitive neuroectodermal tumour, Atrial thrombosis

ABSTRACT

We report three cases of Primitive Neuroectodermal tumor of the Kidney, with inferior vena caval thrombus in two patients and atrial thrombosis in one patient. The role of pre operative diagnosis and chemotherapy needs evaluation in high volume centres.

INTRODUCTION

Renal Primitive Neuroectodermal tumor of the Kidney (PNET) is rare and having more aggressive course towards metastatic disease and death. The five year disease free survival rate for patients presenting with well confined extra-skeletal PNET is around 45-55% and cases with advanced disease at presentation have a median relapse free survival of only two years.[1] The purpose of our study was to review our experience in diagnosis and the management of patients with renal PNET. Review of literature revels that renal PNET is usually cites as a single case report and there is only one series of 16 patients with renal PNET.[2] The importance of preoperative diagnosis, role of neo adjuvant chemotherapy and post operative chemotherapy is unclear and no guidelines are mentioned in literature.

Case History

Case 1

17 year old male patient presented to us with complaints of right sided flank pain and recurrent hematuria on and off since past four months. Investigations revealed it to be a right sided renal mass measuring 10 X 8 cm arising from the upper and mid pole of the right kidney. There was also extension of the tumor thrombus in to the inferior vena cava and extending in to the right atrium. All other metastatic work up was negative.

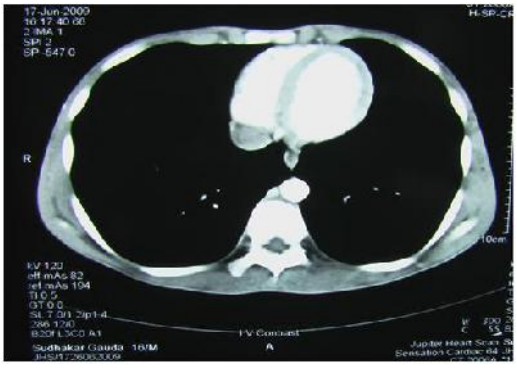

Figure1 CT showing thrombus in right atrium CT showing thrombus in right atrium

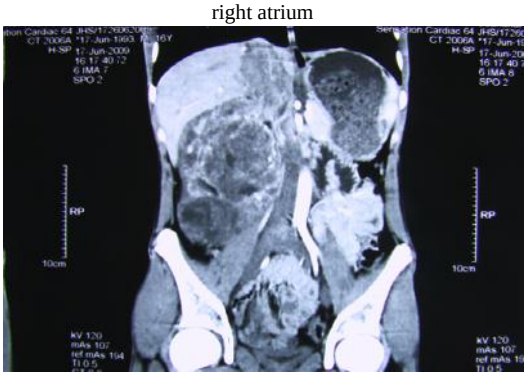

Figure2 CT showing right renal mass

Corresponding author:* **Mhaske S.M
Department of Urology Seth G.S. Medical College Mumbai, Maharashtra, India

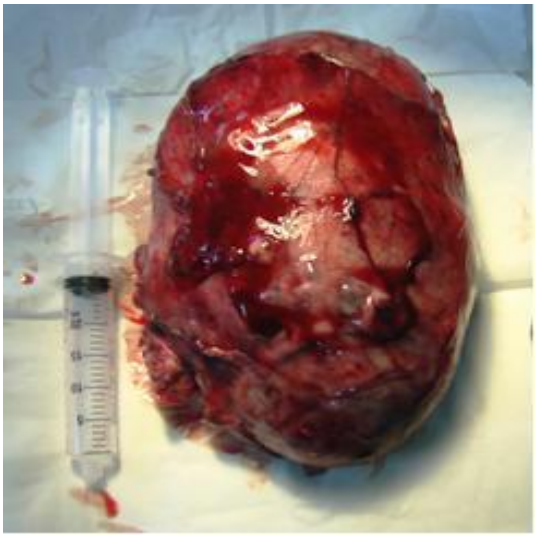

Figure3 Excised specimen of right renal mass.

Right radical nephrectomy, tumour thrombectomy in the inferior vena cava and right atrium, and reconstruction of the inferior vena cava were performed under cardiopulmonary bypass and deep hypothermic circulatory arrest. The patient had an uneventful recovery, but developed sudden onset of breathlessness and a fall in the oxygen saturation on post op day 15. CT pulmonary angiography revealed a large pulmonary embolism of the main pulmonary artery. Immediate anticoagulation was carried out. But within the next two days the patient developed jaundice and enlargement of the left supraclavicular lymph nodes and cannon ball metastasis on X ray chest suggesting a possibility of metastasis. The patient succumbed to his condition on postop day 18.

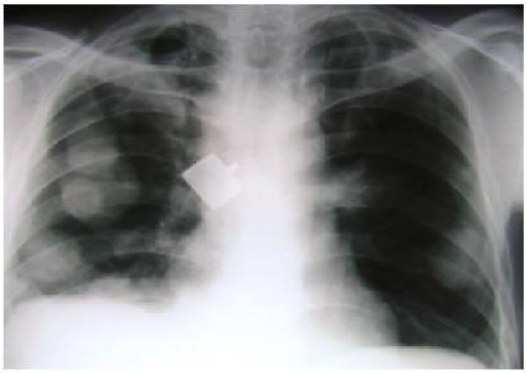

Figure4 Post-op Xray chest showing multiple pulmonary metastasis

Histopathology s/o PNET of kidney.

Case 2

22 year old male with similar complaints had right renal mass with inferior vena cava thrombus.

Right radical nephrectomy with removal of thrombus done. Patient develops pulmonary metastasis postoperatively and died within two months.

Case 3

60 year old male had h/o hematuria and diagnosed to have left renal mass with inferior vena cava thrombus.

Left radical nephrectomy with thrombectomy in inferior vena cava done.

Post op patient received 3 cycles of induction chemotherapy according to RCT-II protocol. Patient was doing well for 2 years but subsequently lost to follow up after that.

DISCUSSION

The clinical signs and symptoms of renal PNET are nonspecific. Preoperative diagnosis of renal PNET cannot be done accurately only with the aid of clinical and radiological examination .Diagnosis is usually based on histopathology and immunohistochemistry postoperatively.[3] None of the cases reported in literature are diagnosed preoperatively.[3456]

Various case reports highlight the high immediate postoperative mortality rate either due to liver or lung metastasis.[5] We too had the same experience with the patient developing pulmonary, liver and supraclavicular metastasis within three weeks. So it is possible that preoperative radiologically insignificant metastasis is missed out or surgical handling of the inferior vena cava thrombus could have led to pulmonary tumour emboli which flare in the post operative immunocompromised stage.

Patients with PNET have certain characteristics. They present at younger age compared to classical RCC and have larger tumour. Inferior vena caval thrombi have been reported in 33% and renal vein thrombi in 49.5% and right atrial in 5 cases highlighting the high incidence of vascular thrombi in PNET.[4] Ours is the 6th reported case with right atrial thrombus.

This in turn reflects on the importance of preoperative diagnosis of PNET. The role of preoperative Fine Needle Aspiration Biopsy (FNAB) and immunohistochemistry probably needs to be revisited in select cases – young patients, unusually large tumour burden and Inferior vena caval thrombi.

CONCLUSION

Due to rarity of tumour and lack of any preoperative clinical and diagnostic examination no randomised prospective studies are possible. Preoperative suspicion is mandatory in young patient with large tumour and inferior vena caval thrombi.

Immunohistoprofile for index cases by preoperative FNAB can be done in large volume centre.

Role of neoadjuvant chemotherapy is refuted in literature but may have a role in select diagnosed cases as many references mention a survival after post operative chemotherapy.[2] Neoadjuvant chemotherapy in large volume centres may be worthwhile.

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How to cite this article:
Mhaske S.M *et al*., Primitive Neuroectodermal Tumour Of Kidney With Right Atrial Thrombosis. *International Journal of Recent Scientific Research Vol. 6, Issue, 6, pp.4407-4409, June, 2015*

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