RENAL CELL CARCINOMA WITH FACIAL SWELLING AND NASAL OBSTRUCTION AS PRIMARY PRESENTATION

Mohd Naqib Mohd Sabri[1], Kasumawati Alli[2], Sze Yin Lam[3], Khairul Azmi Abdul Kadir[3], Norafida Bahari[*1]

[1]Department of Radiology, Hospital Pengajar Universiti Putra Malaysia, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, Serdang, Selangor
[2]Radiology Unit, ParkCity Medical Centre, Kuala Lumpur, Malaysia
[3]Department of Biomedical Imaging, Faculty of Medicine, University of Malaya Medical Centre, Kuala Lumpur Malaysia

***Corresponding author:**
Norafida Bahari, Department of Radiology, Hospital Pengajar Universiti Putra Malaysia, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, Serdang, Selangor.
Email: afidahbahari@upm.edu.my

DOI: https://doi.org/10.32896/cvns.v4n2.25-30
Received: 09.6.2021
Revised: 13.06.2022
Accepted: 22.06.2022
Published: 30.06.2022

ABSTRACT

Background: Renal cell carcinoma (RCC) is a slow growing tumor. About 25–30% of patients are found to have metastases at diagnosis commonly to lung, liver and bones. The incidence of renal cell carcinoma metastasizing to the head and neck has been reported to range from 15-30%. Intranasal mass, or occasionally orbital mass maybe the presenting symptom of metastatic renal cell carcinoma to the nose and sinuses.

Case presentation: We report a case of left RCC with large metastases to the frontonasal region producing head and neck symptoms before the primary lesion could be detected. Clinical presentations of metastatic RCC to the nasal and paranasal regions varies from recurrent epistaxis, nasal obstruction, facial pain, induration or even an orbital mass. In our case, although the patient had typical presentation of metastatic nasal tumour, the diagnosis of metastatic disease was not made.

Conclusion: Patient presented with nasal and paranasal region tumour with no other systemic symptoms, presence of metastatic disease particularly from renal cell carcinoma should be included in diagnosis, as it is a slow growing tumour and the fact that nasal and paranasal areas are the most commonly affected site of metastatic RCC in the head and neck region.

Keywords: Renal neoplasm; Neoplasm metastasis; Paranasal sinuses; Clear cell renal cell carcinoma; Biopsy

INTRODUCTION

Renal cell carcinoma (RCC) is a slow growing tumor. At the time of diagnosis, about 25–30% of patients had metastases (1), most typically to the lung, liver, and bones. Renal cell carcinoma that has spread to the head and neck has been found to occur in 15-30% of cases (2). The presenting symptom of metastatic renal cell carcinoma to the nose and sinuses may be an intranasal or ocular mass.

We present a case of left RCC with extensive frontonasal metastases that caused head and neck symptoms before the main lesion was discovered. The results of ultrasonography, CT, MRI, and angiography imaging are provided.

CASE PRESENTATION

A 70 year-old man presented nine months earlier at another hospital with progressive left-sided nasal obstruction associated with nasal discharge and irregular epistaxis. Contrast- enhanced computed tomography (CT) scan then showed a large expansile

mass occupying the left maxilla with erosion into left orbit, hard palate and extension into the infratemporal fossa. He underwent a medial maxillectomy which revealed a fleshy and vascular intranasal mass. Instead of tumour excision, a biopsy was performed and an initial diagnosis of sinonasal paraganglioma was made. Because MRI was not accessible at the hospital, the patient was referred to our centre for further imaging and care.

MRI of the paranasal sinuses at our centre showed a lobulated mass in the left maxillary sinus, measuring 6.1cm (AP) x 4.5cm (W) x 5.6cm (Ht). The mass returned mixed signal intensity on T1W, T2W and FLAIR with inhomogeneous enhancement in post gadolinium images. Within the structure, there were necrotic zones. The mass extends medially, obliterating the left nasal cavity and causes the nasal septum to deviate to the right. There was bowing of the anterior wall of the maxillary sinus with posterior extension to the anterior wall of the sphenoid sinus (**Figure 1**).

Due to the highly vascular tumour, a pre-operative embolization of the left maxillary tumor was performed revealing that the main feeder artery originated from the left maxillary artery, with feeders originating from the left ophthalmic and right maxillary arteries (**Figure 2**). No arterial supply was demonstrated from both internal carotid arteries (ICA). Endoscopic excision of the left maxillary tumor and ligation of the left external carotid artery was performed. Intraoperatively, the tumor was adherent to the nasal septum and obliterating the entire nasal cavity with erosion of the left orbital floor, lateral nasal floor, lateral wall of the left maxilla and hard palate.

A histopathological examination of the surgical specimen was identified as clear cell carcinoma, most likely arising from differentiated renal cell carcinoma. With the former, which was sinonasal paraganglioma, there were conflicting tissue diagnoses. The 'bane' of this theatrical diagnostic dilemma was eventually determined to be poor initial sample combined with a low suspicion of the tumour being metastatic in nature.

Patient was well post operatively and was planned for radiotherapy at a later date but he defaulted follow-up and was lost to further imaging and treatment.

Patient presented seven months later with persistent left sided nasal obstruction and left facial swelling, as well as a two-week history of right nasal obstruction and epistaxis, at another hospital near to his residence. Repeated contrast enhanced CT scan of the neck showed a heterogeneously enhancing and expansile lobulated mass with necrotic centre measuring 6.0cm (AP) x 6.7cm (W) x 5.8cm (Ht), occupying the entire left maxillary sinus and partial obliteration of the right nasal cavity. There was lateral extension to the left pterygoid fossa infiltrating into the pterygoid muscle, superior extension into the left ethmoidal sinus with bowing of the left lamina papyracea, as well as bowing of the anterior and lateral wall of the left maxillary sinus. Inferiorly it extends to the roof of the oral cavity with posterior extension to the anterior wall of the sphenoid sinus (**Figure 3**).

An ultrasound abdomen later revealed a large heterogeneous and hypervascular solid mass coming from the lower pole of the left kidney, with cystic and hyperechoic components but no calcifications. This was followed by 4-phase CT scan of kidneys, demonstrating a large enhancing exophytic mass with central necrosis in the lower pole of the left kidney, measuring 10.6 x 10.1 x 8.6cm (AP x H x W) in size. The mass extends into perirenal fat but not beyond the Gerota fascia. There were numerous parasitized arteries seen surrounding and supplying this mass. No significant abdominal lymphadenopathy were present and renal vessels were intact. The right kidney was small but no focal lesion within. There were several metastatic nodules seen in the right lung. Hence, this case is a T3aN0M1 renal cell carcinoma or a stage 4 renal cell carcinoma. Patient was referred to clinical oncology department for radiotherapy.

Despite everything that had been stated and done, the patient was unfortunately lost in follow-up and therapy once more.

DISCUSSION

The clinical course and presentation of the renal cell carcinoma (RCC) are variable as it is a slow growing tumor and often go unnoticed until metastasis have occurred (2). The common sites of metastases are lungs, liver and bones (3). In the head and neck region, the nose and the paranasal sinuses is the commonest site for metastasis of RCC (3). However, primary tumour of the nasal and paranasal regions is still the commonest compared to metastatic disease.

Metastatic tumor accounts about 30% of the tumor at this region, based on the report done by Miyahara (4). Bernstein and colleagues did a study on 82 patients with metastatic tumors in the nasal or paranasal sinuses, and found the origin of metastatic tumors was renal cancer in 40 patients (49%), bronchial or lung cancer in 10 patients, breast cancer in 8 patients, testicular cancer in 6 patients, and digestive tract cancer in 5 patients (5).

Clinical presentations of metastatic RCC to the nasal and paranasal regions varies from recurrent epistaxis, nasal obstruction, facial pain, induration or even an orbital mass (6). In our case, although the patient had typical presentation of metastatic nasal tumour, the diagnosis of metastatic disease was not made due to the initial histopathological report which was reported as paraganglioma. Furthermore, patient had no abdominal symptoms to arouse the clinical suspicion of metastatic nasal tumour. From the study done by Ricard S et al. (6), 5 out of 6 patients had presented with unilateral nasal obstruction, which was similarly presented by our patient. They found only 2 patients with epistaxis as the presenting complaint, in contrast with Bernstein JM et al (5) who found epistaxis associated with blood- stained nasal discharge as the most common presenting symptom (70%) of this sinonasal lesion.

Metastatic tumour of RCC at this region is known to be hypervascular (2). As in our case, a hypervascular tumour was well demonstrated on conventional angiogram in which due to its hypervascularity, histopathological sampling was challenging. In another case, as reported by certain writers, RCC can metastasis in the form of an arterio-venous malformation, as Caunter G et al. discovered (10). As a result, a hypervascular tumour is unlikely to be metastatic in origin. As what is usually encountered in RCC, there are mutation in the von Hippel-Lindau (VHL) gene which may lead to abnormal angiogenesis pathway (11) leading to the hypervascular nature of the lesion. According to Yoshimura et al. (7) symptoms of metastatic tumors in the nasal or paranasal sinuses preceded those of primary tumors in 11 out of 18 patients, so when a hemorrhagic tumor is seen in the nasal or paranasal sinuses, renal cancer should be suspected.

The maxillary sinus is the most common site (50%) of paranasal sinuses metastasis from RCC followed by the ethmoidal sinuses, frontal sinuses, nasal cavity and sphenoid sinuses (4). There are several postulations on the route of spread of this metastatic RCC to the head and neck region. Hematogenous spread via systemic circulation was postulated to be through the lungs, leaving microscopic seedings of the lung parenchyma, which would not be visible on the chest radiograph (5). Some postulated these tumours have the ability to bypass the pulmonary capillary filtration mechanism and metastasize directly to the head and neck region via Batson's venous plexus or lymphatic spread via thoracic duct to the head and neck whereby they can anastomose with the great veins of the head or by means of retrograde flow and spread to the nose and sinuses, cutaneous sites and thyroid gland (5,6).

Conflicts arises as there were two histological variances deriving from the same tissue origins as what was observed in our case. However, this is not uncommon due to morphologic similarities between clear cell renal carcinoma and paragangliomas as explained by Lapinski JE et al. (9). For the untrained eye, the morphology and immunohistochemistry of clear cell renal cell carcinoma and paranganglioma are almost identical. However, there are numerous immunohistochemistry markers that might help narrow down the diagnosis; provided there are adequate specimens to be sampled upon. As in our case, when formerly presented with a limited specimen, it ultimately comes down to one's diligence in choosing the best immunohistochemical markers. Extra clinical data such as 'renal mass' as what we had uncovered during ultrasound abdomen when the patient was latterly presented, might surely affect how the tissue diagnosis was made. However, as this case was presented with nasal mass rather than an abdominal mass, we decided to write this case report to emphasise that metastatic renal malignancy should not be overlooked among the many possible tissue diagnosis.

The role of surgery is limited to provide tissue for diagnosis and also surgical debulking of residual disease which may prove very effective after radiotherapy (6). Local symptomatic control with radiotherapy is excellent according to Ricard S et. al (6), whereby they support that the patients with metastatic renal cell carcinoma to the nose and sinuses should be treated with curative intent unless the patient is in the terminal stage of the disease.

CONCLUSION

In conclusion, at this age group of patient who presented to us with nasal and paranasal region tumour with no other systemic symptoms, we should be aware of metastatic disease particularly from renal cell carcinoma as it is a slow growing tumour and the fact that nasal and paranasal areas are the most commonly affected site of metastatic RCC in the head and neck region. For this vascular tumour, pre-embolization of the tumor is excellent to reduce its vascularity as part of pre-surgical management.

STATEMENT OF ETHICS

Informed consents was taken from the patients and each data obtained are treated with extreme confidentiality.

ACKNOWLEDGEMENTS

The authors are grateful to the co-workers and all involved from the Hospital Pengajar UPM(HPUPM), University of Malaya Medical Centre and ParkCity Medical Centre.

CONFLICT OF INTEREST

The authors report no conflicts of interest in this work.

FUNDING

The authors received no funding from any party for this scientific work.

DATA AVAILABILITY STATEMENT

The authors are contactable via emails for further enquiries.

REFERENCES

1. Nayak DR, Pujary K, Ramnani S, Shetty C, Parul P. Metastatic renal cell carcinoma presenting with epistaxis. Indian Journal of Otolaryngology and Head and Neck Surgery. 2006 Oct;58(4):406-8.
2. Ricard S, Andrew JS, Simon PH, Patrick RA, Andrew RB, Nick JS, W Trevor Farrington. Metastatic renal cell carcinoma to the nose and paranasal sinuses. Head Neck. 2000 Jan; 22: 722–727.
3. Homer JJ, Jones NS. Renal cell carcinoma presenting as a solitary paranasal sinus metastasis. J Laryngol Otol 1995;109:986–989.
4. Tunio MA, Hashmi A, Rafi M. Epistaxis and Proptosis - Unusual primary manifestations of metastatic renal cell carcinoma. Pak J Med Sci 2009;25(6):1012-1014.
5. Matsumoto Y, Yanagilhara N. (1982): Renal cell carcinoma metastatic to the nose and paranasal sinuses. Laryngoscope 92:1190-1193. .
6. Yoshimura T, Suenaga T, Kozaki H, Oohashi N, Sakai N. A case of renal cell carcinoma revealed by metastasis to the nasal cavity and paranasal sinuses. Sapporo Hosp Med 1989; 49: 31–5.
7. Miyahara H. Metastastic carcinoma to head and neck lesion. J Otolaryngol Jpn 1983; 86: 951–7.
8. Bernstein JM, Montgomery MD, Balogh K Jr. Metastatic tumors to maxilla, nose, and paranasal sinuses. Laryngoscope 1966; 76: 621–50.
9. Lapinski JE, Chen L, Zhou M. Distinguishing clear cell renal cell carcinoma, retroperitoneal paraganglioma, and adrenal cortical lesions on limited biopsy material: utility of immunohistochemical markers. Appl Immunohistochem Mol Morphol. 2010 Oct;18(5):414-21.
10. Caunter G, Faeez Md Noh MS, Safri LS, Kumar K, Md Idris MA, Harunarashid H, Yahaya A. Delayed Presentation of Metastatic Renal Cell Carcinoma as an Arteriovenous Malformation Mimicking Vascular Tumour of the Forearm. EJVES Short Rep. 2019 Jul 31;44:19-22.
11. Albandar HJ, Roberto ES, See JR and Sabiers JH: Arteriovenous malformation and thyroid metastasis from underlying renal cell carcinoma, an unusual presentation of malignancy: A case report. Oncol Lett 13: 3323-3327, 2017.

FIGURE LEGENDS

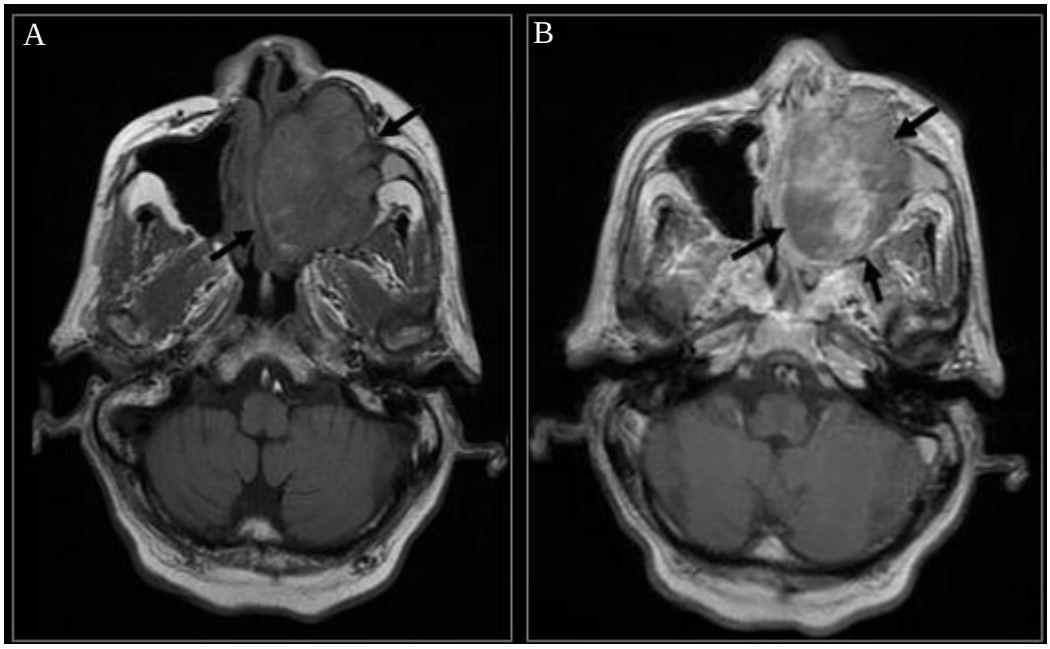

Figure 1: Axial spin-echo T1-weighted MR (A) pre-gadolinium and (B) post-gadolinium images of the nasal cavity and maxillary antrum (B) showing tumor mass in the left maxillary antrum and nasal cavity (black arrows) returns mixed signal intensity and inhomogeneous enhancement. This mass extends medially into the left nasal cavity causing obstruction of the left nasal cavity and deviation of nasal septum to the right. Anteriorly it causes bowing of the anterior wall of the maxillary sinus.

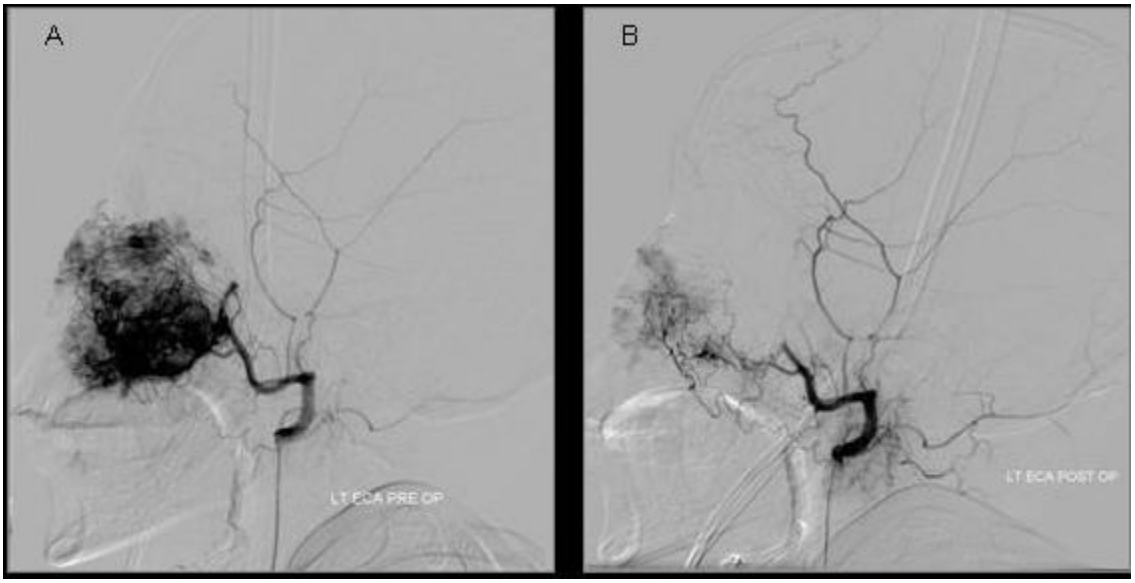

Figure 2: Digitally-subtracted left external carotid angiogram in lateral view during (A) pre-embolization of the left maxillary tumour which showed a hypervascular mass with the main feeder artery arising from the left maxillary artery. (B) Post-embolization showed marked reduction in the vascularity of the tumour.

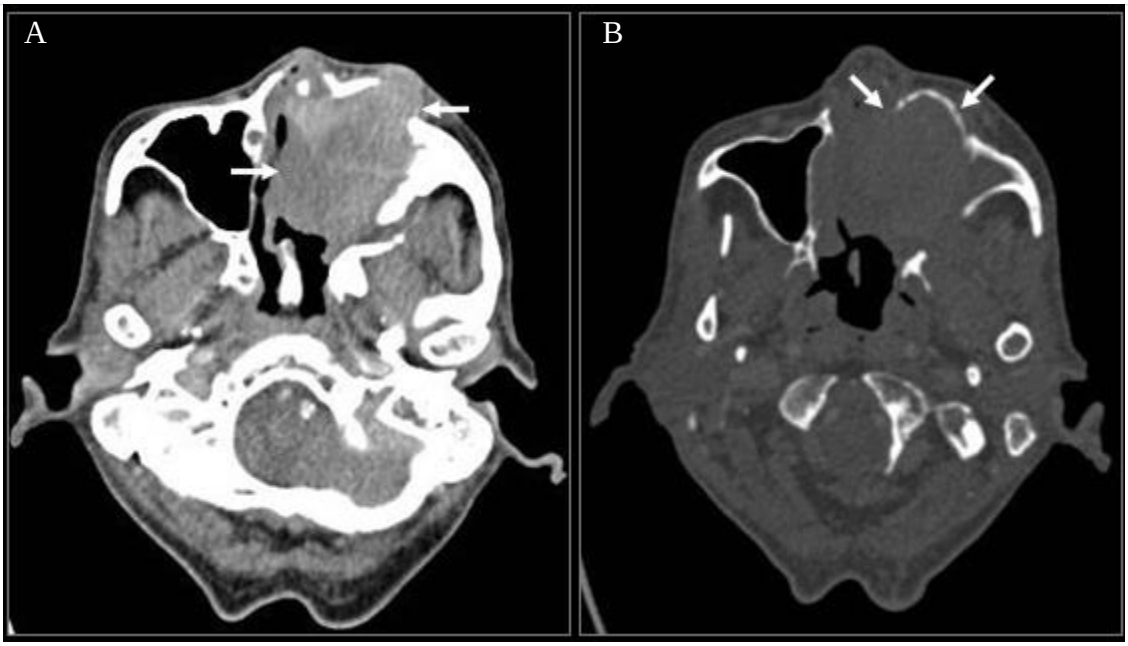

Figure 3: Axial images from contrast-enhanced CT of head and neck in (A) soft tissue window and (B) bone window demonstrating the left maxillary tumor (black arrows) with extensive local infiltration and destruction.

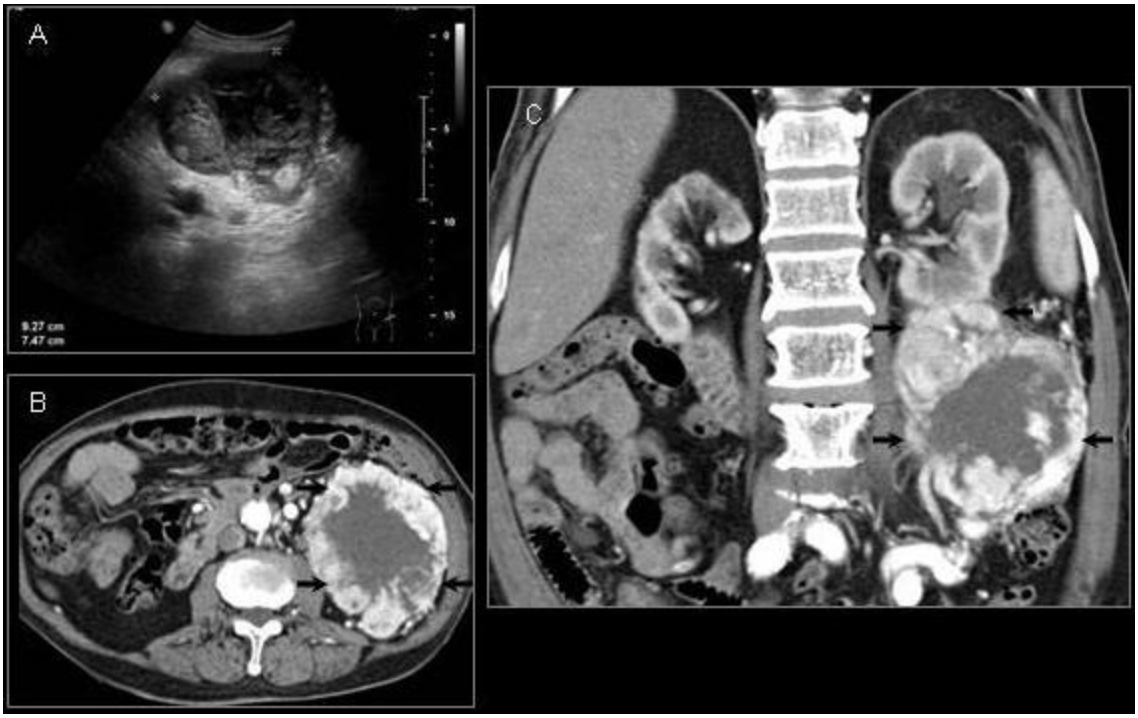

Figure 4: Left para-sagittal ultrasound image (A) showed large heterogenous tumour in the lower pole of left kidney. Contrast-enhanced CT scan of the abdomen in (B) axial plane and (C) coronal reformatted image showing a hypervascular left renal mass (black arrows) with necrotic centre and multiple parasitized arteries supplying the mass.