Isolated cauda equina metastasis from renal cell carcinoma – A rare cause of intradural-extramedullary compression

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Abstract
Intradural extramedullary spinal metastasis is rare, representing 6% of all spinal metastasis. Indeed, intradural metastasis from a Renal Cell Carcinoma to the cauda equina is extremely rare with only 11 case reports present in the past. We present a patient with Cauda equina syndrome with an intradural extramedullary lesion causing compression of the nerve roots. He was subjected to a surgical decompression of the cauda equina with excision of the mass. The pathological examination displayed metastatic clear cell RCC with infiltration of the cauda equina. Thus, metastatic tumors constitute an important differential diagnosis for all lesions of the spine irrespective of level or location.

Keywords: Metastatic, Renal cell carcinoma, Intradural, Extramedullary, Cauda equina.

Introduction
Renal cell carcinoma accounts approximately for 85% of all renal tumors and 75% are of the clear cell type.[1] Renal Cell Carcinoma (RCC) has the highest mortality rate of the genitourinary cancers and the incidence of RCC has risen steadily. In recent decades, the incidence of RCC has been steadily rising by 2–4% each year and RCC is now the 7th leading cancer type in men.[9] Like most common cancers, RCC occurs in either a sporadic or inherited pattern. Thirty percent of patients with RCC have metastasis at presentation, and 30% to 50% of patients who undergo radical excision of the lesion.[13]

The majorities of cauda equina tumors are of glial or nerve sheath origin[3,5,7,15] and metastases from outside the central nervous system are extremely rare. To our knowledge, only 11 cases (RCC) with metastatic tumor of the cauda equina from outside the central nervous system have been reported in detail in literature.[1,2,4,6,8,12,14] The literature is reviewed with reference to tumor pathologies, clinical findings and route of metastasis to the cauda equina.

Case Report
A 54 year old male presented to the outpatient clinic with inability to walk or stand gradually progressing for 1 year with acute worsenning of 3 days. He was a known case of cerebrovascular accident with right hemiparesis managed with medical therpay as well as a known case of renal cell carcinoma of the left kidney for which he underwent a left partial nephrectomy a year ago. The local disease was under control for a year with regaular surveillence. He also had a right non-functional kidney due to chronic kidney disease with diabetes mellitus and essential hypertension as well.

Preoperative MRI was done and was found to have a well defined rectangular shaped focal L3-4 intradural extramedullary lesion 3.2* 1.9*1.4 mm with displacement of cauda equina. The mass was well demarcated and demonstrated homogenous enhancement. (Fig. 1, and 2)

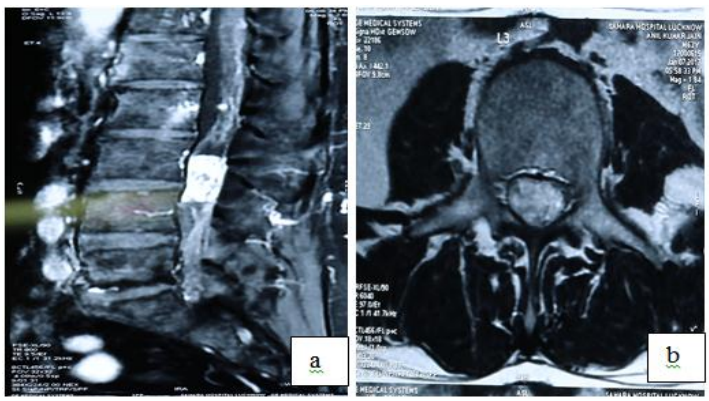

Fig. 1(a) and (b): MRI of the Lumbosacral Spine- T1 with contrast sequence shows an intradural extramedullary lesion at the level of L3-L4 occupying the entire spinal canal.

Abdominal MR images and radioisotope images revealed no tumor recurrence of the primary lesion and no tumor invasion to the intrapelvic or paraspinal organs. (Fig. 3, and 4) The thoracic cord, cervical cord and brain MRI scans showed no evidence of tumor.

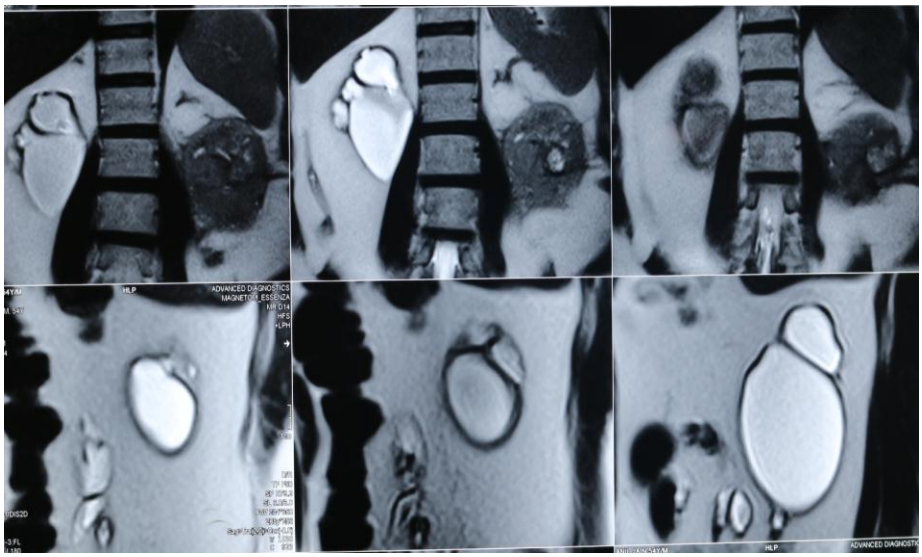

Fig. 2: MRI of the abdomen showing a grossly hydronehrotic left renal pelvis with a resected small right partially functional kidney

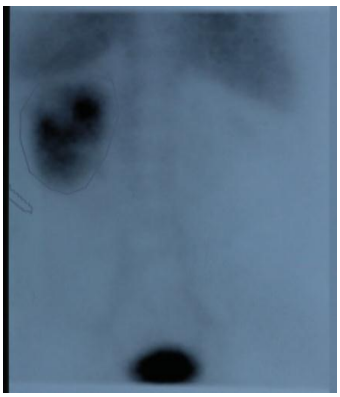

Fig. 3: Radionucleotide imaging showing no metastasis with only one functioning kidney.

The patient underwent L3 laminotomy and excision of the lesion with decompression of the cauda equina.

On table, the dura appeared bulged and was dark in colour. On opening the thickenned dura there was profuse bleeding. The tumor was large, diffuse and interspersed with nerve roots. There were no obvious feeding arteries oar draining veins visible over the tumor. The tumor was not encapsulated, firm, friable and highly vascular. Thus the tumor had to be removed in pieces with proper neurovascular monitoring. In view of the unprecedented vascularity as well as the unusual fofrm of the tumor a Frozen section pathology examination was requested. This frozen section was reported as haemangiopericytoma of the spine, which further complicated the issue presenting an additional feature and diagnosis of the already complicated slate.

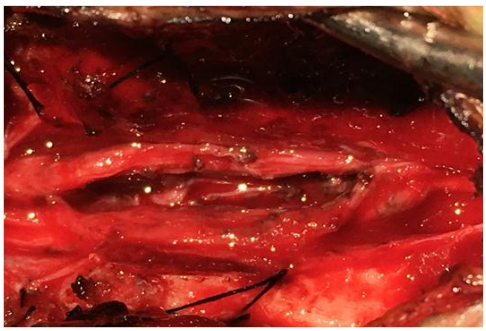

Fig. 4: Intraoperative image of the open thickenned dura at L3-L4 showing profuse bleeding and a decompressed spinal canal

Post procedure, the patient neurological status remained static but he regressed nephrologically due to the stress of surgery combined with the progression of the diabetes and hypertension. He became dialysis dependent and eventually went into urosepsis. Hence despite the final diagnosis being known it wasn't possible to subject him to chemotherpy. He was treated with antibiotics in intensiv care, cleared of the infection and discharged to review periodically for haemodialysis and nephrological monitoring.

The final Histopathological analysis was consistant with features of metastatic renal cell carcinoma. It showed Tumor cells arranged in alveolar pattern with prominent delicate vasculature separating the tumor lobules. Tumor cells show small nuclei with clear

cytoplasm. Immunohistochemistry for pan CK and CD10 is positive in tumor cells and negative for chromogranin and synaptophysin. Confirming the diagnosis.

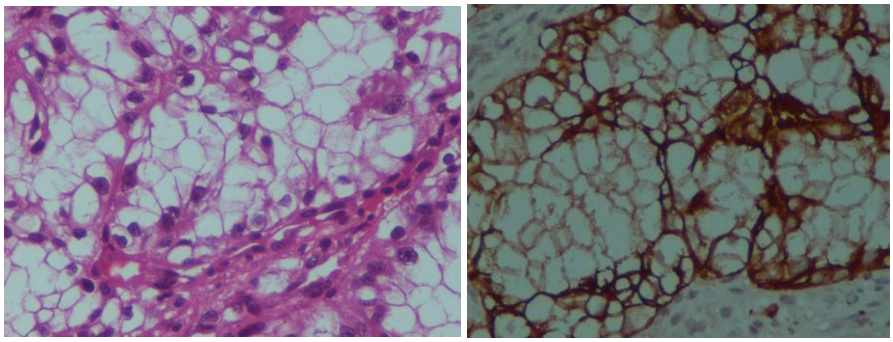

Fig. 5: Histopathological slide of the tumor showing the alveolar pattern of cellular arrangement with cells showing small nuclei and clear cytoplasm. Special stains show a loss of synaptophysin characteristic of Renal Cell Carcinoma

A PET CT scan was advised to look for other foci of metastasis, but showed no other loation of spread. We also attempted to look for pathways to delineate the possible pathophysiological mechanism of the spread of this tumor. No clear mechanism was revealed however.

Patient was discharged from the hospital and is being followed up regularly.

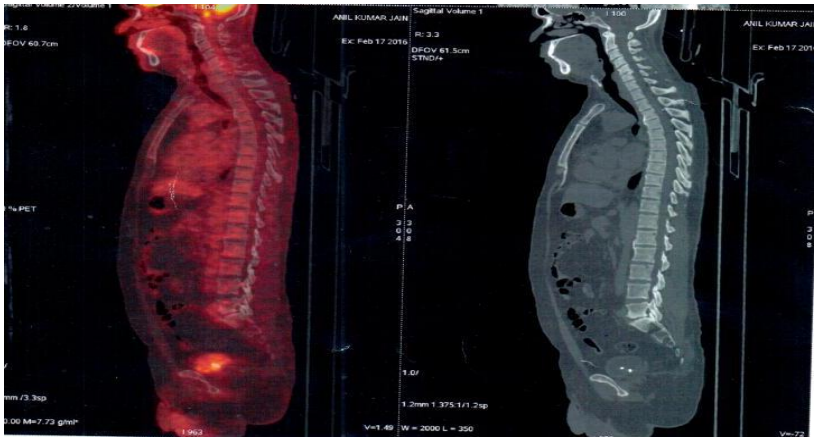

Fig. 6: PET images post surgery showing no other foci of spread

Discussion

RCC shows a high tendency to metastasize, with 30% of patients having metastasis at presentation and 30% to 50% of patients undergoing radical excision.[1] In this report, the authors report a patient with RCC metastasizing to the cauda equina and discuss possible mechanisms of intradural metastatic spread without bone involvement. Increased understanding of the spreading of this pathology offers opportunities for the improvement in medical and surgical techniques.

Five routes have been hypothesized for metastatic intradural spinal tumor from outside the central nervous system;[1] (a) haematogenous, via the arterial system, (b) through the rich venous plexus (Bateson's venous plexus), (c) through perineural lymphatics, (d) spread via subarachnoid space, (e) seeding from the involved osseous structure to the cerebrospinal fluid through the dura mater, and (f) Arterial embolism.

To explain the unusual metastasis to the cauda equina. Some authors have suggested venous embolism through the venous channels between the pelvis and the spinal cord. Others have proposed that seeding via the subarachnoid space formed the metastasis around the cauda equina as drop metastasis[10,11] This seems unlikely because brain MRI showed no metastatic brain tumor and his surgical findings revealed no evidence of subarachnoid dissemination of the tumor. Previous reports displayed that 50% of brain metastases of RCC occur in the cerebral cortex, 10% in the cerebellum, 8% in the meninges, 2% in the pituitary gland, and 1% in the brain stem.

The mechanism of the spread of metastatic disease to the cauda equina can be understand, as previously reported, via the rich venous Batsons plexuses and the perineural lymphatic vessels, through the subarachnoidal space, or by direct extension of the primary tumor. A combination of these mechanisms

may be the pathophysiological pattern in our unusual case. The demonstrated absence of other spinal metastases may reduce the likelihood of systemic hematogenous spread.

Despite a radiological work-up, we could not diagnose correctly before surgery. As the lesion resembled a typical intradural extramedullary lesion such as a schwannoma or a meningeoma it was reported as such without suspicion of other diagnoses.

Conclusion

The majority of cauda equina tumors are primary tumors, and metastases are very rare. However the presence of a long standing primary malignancy with the emergence of a space occupying lesion elsewhere must strongly arouse suspicion of a metastasis despite the apparently aberent nature of the spread and the supposed controlled nature of the primary lesion.

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