Unusual association of diseases/symptoms

The occult nature of intramedullary spinal cord metastases from renal cell carcinoma

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Summary

Renal cell carcinomas (RCC) are characterised by a tendency to metastasise widely, often while remaining occult. Intramedullary spinal cord metastases (ISCM) from RCC may be the presenting feature of the disease or present at any time in the disease course. This case report discusses an ISCM from RCC which became manifested at the time of resection of the primary tumour. We review the literature published on ISCM from RCC from 1990 to date comparing disease characteristics and presentations.

BACKGROUND

A 62-year-old male smoker with no significant medical history or known genetic syndrome was referred by a peripheral hospital to the urology service at our institution with a 4-week history of worsening right flank pain associated with haematuria and urinary frequency. A CT scan of the thorax, abdomen and pelvis demonstrated a large heterogeneous mass in the right kidney with perinephric lymphadenopathy. Moreover, multiple rounded lesions throughout both lung fields were noted to be consistent with metastases. Brain imaging did not reveal a mass lesion. A right radical nephrectomy was performed and the patient was discharged home 6 days later. Histopathological evaluation of the nephrectomy showed clear cell renal cell carcinoma (RCC) located in the superior pole of the right kidney, 144 mm×92 mm×115 mm in dimension. It extended into the renal vein and invaded the renal sinus. Retro-caval lymph node involvement was also confirmed on histology.

CASE PRESENTATION

Seventeen days after the surgery, this gentleman was presented to his local hospital complaining of severe back pain, progressive lower limb weakness and urinary incontinence. Physical examination demonstrated dense paraparesis (Medical Research Council (MRC) grade 1 of 5) without sensory deficit.

INVESTIGATIONS

MRI of the cervical spine (figure 1A,B) showed a syrinx with an enhancing homogenous intramedullary lesion measuring 7 mm×12 mm×8.5 mm at the level of C7. The rest of the MRI whole spine with gadolinium was normal with no evidence of enhancing area on the spinal cord or vertebral area.

DIFFERENTIAL DIAGNOSIS

Primary intramedullary tumour.

TREATMENT

The patient underwent a C7 laminectomy with surgical resection of intramedullary lesion, which proved pathologically to be metastatic carcinoma with morphology and immunophenotype in keeping with metastatic RCC (figure 1C,D). During the postoperative period, his lower limb weakness improved to MRC grade 3 of 5.

OUTCOME AND FOLLOW-UP

He was transferred back to the referring hospital for palliative treatment. The patient passed away 3 months later due to progression of disease.

DISCUSSION

RCC accounts for 2–3% of all malignancies¹ with a tendency to spread haematogenously. Sites frequently affected by metastatic RCC include the lungs (50% of cases), bone (49% of cases)² and the central nervous system (CNS). Roughly one-third of patients with RCC have metastatic disease at the time of diagnosis.³ Moreover, RCC accounts for 4% of Intramedullary spinal cord metastases (ISCM),⁴ which have a relatively poor prognosis when compared with vertebral metastases.⁵ Thus, progressive neurological symptoms in patients with RCC should be recognised and investigated urgently as early intervention is crucial. However, documented clinical features of patients with ISCM are variable.⁶

ISCM from RCC was first reported by Von Pfungen in 1906.⁷ Schijns *et al*⁸ have reported an ISCM as the initial manifestation of RCC. MRI in contrast is regarded as the most sensitive imaging investigation for the identification and localisation of spinal metastases^{9–11} and can aid in early detection of these lesions. Any patient with ISCM should also have brain imaging as multiple metastatic lesions may occur concurrently in the CNS and occur six times more frequently in ISCM compared with metastases that involve the spine.⁵ Positron emission tomography may help to discriminate diagnostically equivocal lesions detected by imaging.¹²

Figure 1 (A) T1-weighted sagittal MRI with gadolinium showing an intramedullary mass lesion at the C7 level. (B) T1-weighted axial MRI with gadolinium showing an intramedullary mass lesion at the C7 level. (C) Photomicrograph showing metastatic renal cell carcinoma (RCC). (D) Photomicrograph showing metastatic carcinoma. CD10 immunohistochemistry showing positive labelling (brown signal) of cytoplasm and cell membranes in keeping with RCC (clear cell type). CD10 immunostain $\times 200$.

Table 1 Characteristic of patients with intramedullary spinal cord metastasis (ISCM) secondary to renal cell carcinoma (RCC)

Author	Number of patients	Time from diagnosis of RCC to ISCM development			Level of ISCM Metastasis			Concomitant brain metastases
		Concomitant	<1 year	>1 year	Cervical	Thoracic	Lumbar	
Ateaque <i>et al</i> ¹³	1	–	–	1	1	–	–	–
Schijns <i>et al</i> ⁶	1	1	–	–	–	1	–	–
Fakih <i>et al</i> ¹⁴	6	2	2	2	3	2	1	3
Poggi <i>et al</i> ¹²	1	–	–	1	–	1	–	–
Kaya <i>et al</i> ¹⁶	1	–	–	1	–	–	1	–
Gomez de la Riva <i>et al</i> ¹⁷	1	1	–	–	–	–	1	–
Donovan <i>et al</i> ¹⁸	1	–	1	–	–	1	–	–
Asadi <i>et al</i> ¹⁹	1	1	–	–	–	–	1	1
Total	13	5	3	5	4	5	4	4

Table 2 Most common clinical manifestations of intramedullary spinal cord metastasis (ISCM) from 12 patients with known renal cell carcinoma (RCC)

Symptoms at initial presentation	Number of patients
Upper limbs weakness	1
Lower limbs weakness	8
Bilateral limbs weakness	1
Urinary incontinence	4
Spinal tenderness	3
Brown-Sequard syndrome	3

We identified eight publications reporting ISCM from RCC from the year 1990 to date. Table 1 compares the reported patient characteristics. Of 13 patients, five had concomitant ISCM at initial diagnosis of RCC. Two patients developed an ISCM 11 years after the diagnosis of RCC, highlighting the diverse nature of tumour behaviour.^{13 14} The most common location for ISCM from RCC was the thoracic cord (five patients). Four patients were also found to have brain metastases at the time of diagnosis, in contrast to our patient where brain imaging did not show evidence of metastatic tumour.

Table 2 demonstrates the most common clinical manifestations of ISCM from RCC, with lower limb weakness the most frequent finding (8 of 12 patients). A total of 80% of patients with vertebral metastases and spinal cord compression from metastatic RCC complain of pain at earliest presentation; in contrast, spinal tenderness was present in 3 of 12 patients with ISCM.¹⁴

The prognosis of patients with ISCM is poor, and treatment modalities vary depending on patients age, clinical findings and Eastern Cooperative Oncology Group status. Moreover, RCC is regarded as a relatively radioresistant tumour. Therapeutic or palliative options include conservative management, radiation therapy or surgical resection. The median survival of patients who receive radiotherapy is 4 and 2 months for those not receiving radiotherapy.⁶ The use of stereotactic radiosurgery has become more widespread due to recent advances in the treatment of extracranial lesions.¹⁵

Learning points

- We highlight the rarity of the presentation of intramedullary spinal cord metastases (ISCM) from renal cell carcinomas (RCC). Review of the literature suggests that due to the relative occult nature of RCC, ISCM from RCC may present at the same time of the initial diagnosis or many years later.
- We highlight the presence of central nervous system metastases which include ISCM and emphasise the importance of clinical awareness in patients who presented with neurological deterioration.
- MRI of the spine in contrast is indicated in symptomatic patients to rule out any evidence of metastatic disease.

Competing interests None.

Patient consent Obtained.

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