Unusual association of diseases/symptoms

Renal cell carcinoma metastasis to the paranasal sinuses and orbit

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The authors report a rare case of renal cell carcinoma (RCC) metastasis to the paranasal sinuses. The authors review RCC and its potential for sinonasal metastasis and discuss the variable presentation and need for clinical suspicion for early diagnosis and treatment. A 74-year-old man presented with numbness of the left side of the face, reduced visual acuity and ptosis 12 years after nephrectomy for RCC. Imaging studies showed a lesion in the left pterygopalatine fossa and the histological features supported the diagnosis of metastatic RCC. RCC metastasis to the paranasal sinuses is very rare and can present with various symptoms depending on the affected organ. These symptoms occasionally are the initial manifestation of renal RCC and it is very important to recognise them so that the patient receives the appropriate therapy to improve survival.

BACKGROUND

Renal cell carcinoma (RCC) is the commonest malignant renal tumour, most commonly affecting men between the ages of 30 and 60 years.¹ RCC is a tumour with a very unpredictable course.² RCC metastases to the sinonasal area are rare with only a few reported cases in the English-language literature.

We present the case of RCC metastasis to the paranasal sinuses and orbit 12 years after treatment for the primary tumour and discuss the different manifestations of metastases of RCC to the sinonasal area.

CASE PRESENTATION

A 74-year-old man was referred to ophthalmology with a 6 month history of numbness of the left side of the face, reduced visual acuity and ptosis. His medical history included RCC in 1998 treated with nephrectomy and a right eye enucleation for melanoma in 1967.

On examination, the patient had swelling and erythema of the left side of his face, right sided enucleation, ptosis of the remaining left eye and left third and sixth cranial nerve palsy. His visual acuity in the remaining eye was on admission 6/60. Because of the rapid deterioration of the patient's visual acuity he was commenced on intravenous steroids and his visual acuity was improved to 6/24.

INVESTIGATIONS

An urgent MRI scan showed a lobulated mass extending from the left pterygopalatine fossa superiorly and medially into the orbit through the inferior orbital fissure causing proptosis with involvement of the posterior ethmoid air cells and the cavernous sinus. The mass was displacing the optic nerve superiorly within the posterior segment of the orbit (figure 1). The patient was referred to the Otolaryngology Department.

Before endonasal biopsies and orbital decompression a CT-scan was performed again showing a mass in the left pterygopalatine fossa that extended into the surrounding bony structures (figure 2).

During the operation there was persistent bleeding from the biopsy sites which was managed with packing and placement of silastic sheet over surgical areas.

Histology showed tumour composed of cells with clear cytoplasm arranged in glandular and solid architectural patterns, surrounded by a rich vascular network. There was mild pleomorphism (figure 3). The cells showed strong

Figure 1 MRI scan showing a lobulated mass in the left pterygopalatine fossa extending into the orbit producing proptosis.

Figure 2 CT scan showing a mass in the left pterygopalatine fossa with destruction of the left greater wing of the sphenoid and erosion of the lateral wall of the left sphenoidal sinus.

immunochemical positivity for CD10 and Vimentin, with focal positivity for CK19, CK 8/18, S100 and Ca 19.9 (figure 4). The overall morphological features and immunochemical profile supported the diagnosis of metastatic clear cell carcinoma from a renal clear cell carcinoma primary site of origin.

A CT-scan of the chest, abdomen and pelvis only revealed a small subcutaneous nodule measuring 10 mm on the right side of the sternum and an 11 mm nodule in the thymic fat which were suspicious of metastasis.

TREATMENT

Following discussion in the head and neck multi-disciplinary meeting palliative radiotherapy was offered to the patient.

DISCUSSION

Differential diagnosis of sinonasal pathology is vast. Diagnosis and treatment of malignancies requires early clinical suspicion to prompt imaging and histology. Our case demonstrates symptoms and signs of malignancy but also highlights that even a distant history of RCC and features of vascular dissection intraoperatively are important pointers to RCC involvement.

Sinonasal RCC mets are rare and share presenting signs and symptoms with other pathologies particularly other tumours.

Metastatic tumours to the head and neck region from infraclavicular primaries are infrequent and metastatic disease in the paranasal sinuses is rare. The commonest neoplasms to metastasise to the head and neck region are breast and lung carcinoma followed by RCC.^{3 4} The commonest metastatic tumour to the paranasal sinuses is RCC, accounting for 49% of the cases, followed in order of frequency by bronchus, urogenital ridge, breast and gastrointestinal tract. In many cases the symptoms of sinonasal metastatic disease are the initial presenting symptoms of

Figure 3 Metastatic renal cell carcinoma with cells containing clear cytoplasm arranged in glandular and solid architectural patterns (H&E stain).

the primary tumour. Metastatic tumours in the paranasal sinuses are difficult to differentiate from primary tumours in the same location both clinically and radiologically and it is only with biopsy that the true nature of the tumour is revealed.⁵

RCC (hypernephroma, adenocarcinoma of renal parenchyma) comprises approximately 85% of all malignant renal tumours and typically occurs in men between the ages of 30 and 60 years.¹ Characteristically, the tumour is slow-growing and encapsulated in its early stages and therefore may be asymptomatic until late stages.⁶ The initial classical presenting triad of haematuria, costovertebral pain and abdominal mass only occurs in 9% of the patients and in some cases sinonasal metastasis is the presenting feature.³ The clinical course of the primary tumours is often unpredictable. They vary in their growth rate, metastatic potential and patterns of metastatic spread. Spontaneous regression has been noted, as has the occurrence of metastasis several years following treatment of the primary,² with reported cases of metastasis from RCC up to 31 years after nephrectomy.⁷ In a study by McNichols *et al*, which analysed the survival of 506 patients with RCC, 11% of the patients had recurrence 10 years after nephrectomy.⁷

Metastasis are present at the initial diagnosis in 25–33% of the patients.⁸ Synchronous and metachronous metastasis are usually multiple but in 1.6–3.2% of cases solitary (1.5% after nephrectomy).⁸ The most common sites of metastasis are the lungs (75% of cases), regional lymph nodes (65%), bone (40%) and liver (40%).² Approximately 15% of patients with RCC have extracranial head and neck metastases.¹ Metastatic RCC is macroscopically rich in vascular components elastic, soft, yellowish white in colour tumour^{9 10} and its histopathologic characteristics include the presence of encapsulating connective tissue, clear cell borders, round or oval nuclei and abundant clear cytoplasm.^{10 11} Metastatic tumours originating from renal cancer are vascular in nature^{2 9–11} and as in our case significant haemorrhage should be expected. RCC is frequently noted to invade the local vascular network by direct extension into the interlobular, arcuate and interlobar veins and metastasises through the systemic circulation. In an attempt to explain the occasional presentation of head and

Figure 4 Metastatic renal cell carcinoma (RCC) with cells showing strong CD-10 immunochemical positivity, characteristic for RCC.

neck mass without evidence of metastatic disease elsewhere many theories have been developed such as spread via Batson's venous plexus or lymphatic spread via the thoracic duct.¹

Signs and symptoms produced by the metastases are dependent on the affected organ and local invasion from the metastases.¹ These features are occasionally the initial presentation of the RCC and it is very important to recognise them so that the patient be managed appropriately to improve survival. We conducted a Medline literature review for sinonasal RCC metastasis. In particular we looked at the initial symptoms and the time of presentation. Our search strategy, shown in appendix 1, resulted in 28 cases reported in 19 English language papers.^{1–5 8–21}

The commonest presenting complaint (table 1) was epistaxis (46.4%), followed by nasal obstruction (35.7%). The symptoms caused by the metastatic tumour presented after the diagnosis and treatment of the primary RCC in 60.7% of the patients (with interval from 4 weeks up to 17 years after treatment for the primary tumour) and in 39.3% of the patients the metastatic disease was the first manifestation of the RCC.

The 5-year survival rate after a nephrectomy is approximately 60% to 75%. In those patients who have a solitary metastasis, the treatment is radical excision and nephrectomy, with an approximate 5-year survival of 35%, however the prognosis of patients with multiple metastases is poor, with the 5-year survival being 0% to 7%.^{11 13}

Table 1 Presenting symptoms of metastasis to the sinonasal area

Presenting symptoms		
Presenting complaint	Frequency	Percent
Proptosis	3	12
Epistaxis	11	44
Facial swelling	7	28
Visual loss	2	8
Nasal obstruction	9	36
Frontal headache	2	8
Diplopia	4	16
Epiphora	1	4

Treatment should be individualised according to the tumour location and the general health of the patient. Surgery is probably the optimal treatment for patients with single sinonasal metastasis but for irresectable tumours there are other available therapies, which include RT, chemotherapy and immunotherapy.⁴ Immunotherapy has been shown to have the potential for cure even in metastatic RCC, depending on the patients' general health. Chemotherapy is now generally reserved for patients in whom immunotherapy has failed.^{4 11}

Learning points

- ▶ Metastatic tumours to the sinonasal area are very rare.
- ▶ RCC is the commonest metastatic tumour to the paranasal sinuses.
- ▶ This paper describes a case of RCC metastasis to the paranasal sinuses and orbit.
- ▶ Sinonasal metastases present with specific signs and symptoms which should alert the clinician.

Competing interests None.

Patient consent Obtained.

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