The Journal of Laryngology and Otology
October 1995, Vol. **109,** pp. 986–989

Renal cell carcinoma presenting as a solitary paranasal sinus metastasis

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Abstract
We present an unusual case of renal cell carcinoma presenting as a periorbital mass. The presenting symptom was a supraorbital swelling which proved to be a mucocoele which had arisen secondary to a solitary metastasis in the ethmoidal sinuses. We discuss the management strategy including the place of endoscopic biopsy, the choice of investigations and craniofacial resection.

Key words: Renal neoplasm; Neoplasm metastasis; Paranasal sinuses; Nose; Mucocoele; Endoscopy; Biopsy

Introduction

Metastatic disease in the nasal cavity and paranasal sinuses is rare. Although renal cell carcinoma is the commonest metastasis in this area (Nahum and Bailey, 1963), metastases in this disease are seldom solitary (1.6–3.6 per cent: Middleton, 1967) and such metastases usually present after the primary tumour has been diagnosed. Furthermore, very few cases have been reported where the metastasis has manifested itself as a periorbital mass (Wolin, 1993).

Case report

A 59-year-old female was referred to the Ophthalmology Department with a three-week history of a right periorbital mass increasing in size and causing diplopia and discomfort. She was otherwise fit and well. On examination, she was found to have a 2 cm mass in the right medial aspect of her supraorbital margin (Figure 1). A CT scan showed a soft tissue mass occupying the right ethmoidal air cells extending to the right frontal sinus and into the orbit (Figure 2). It was thought most likely to be a mucocoele, except for the presence of some flecks of calcification at the superior margin of the lesion.

She was referred to the ENT Department and, at endoscopy, the mass was found to extend into the lateral nasal wall. This was biopsied endoscopically at which time there was profuse bleeding which was controlled by packing the lesion with surgicel. The mass was magenta in colour and contained speckles of bone within it.

Post-operatively, a carotid angiogram showed a high degree of vascularity with feeding vessels mainly from the right ophthalmic artery but also from the left ophthalmic artery and right maxillary artery (Figure 3). Histology showed the mass to be a clear cell carcinoma (Figure 4). A subsequent ultrasound scan of her abdomen showed a left renal mass 4 cm in diameter, in the region of the upper pole.

A CT scan of her abdomen and thorax confirmed this finding and showed no other metastases. A left nephrectomy was carried out by the urologists, at which the tumour was found to extend to, but not breach, the capsule and two satellite deposits in the same kidney were noted. Histology showed clear cell carcinoma with no capsular breach nor lymphatic or hilar vessel involvement. She made an excellent recovery.

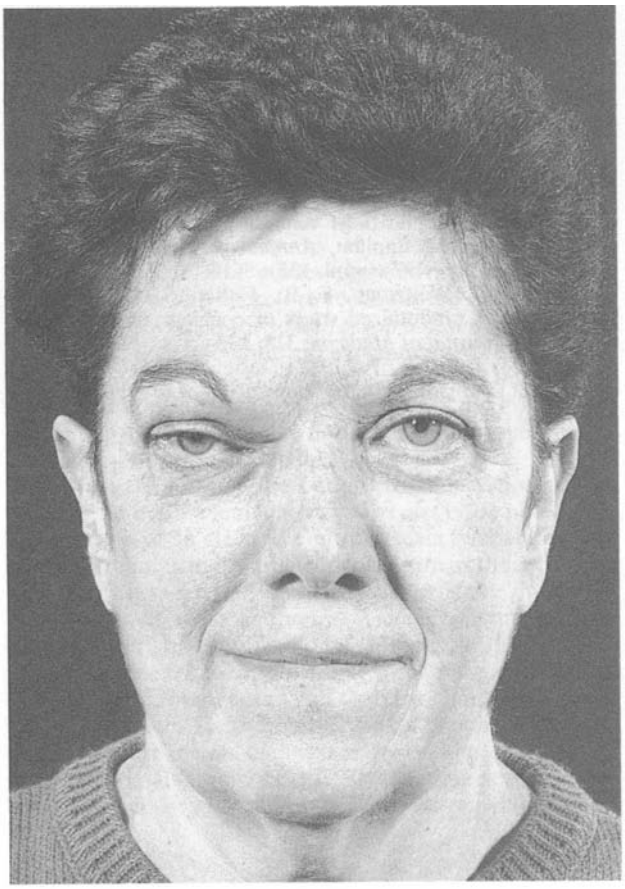

FIG. 1
Pre-operative photograph of patient.

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Accepted for publication: 1 July 1995.

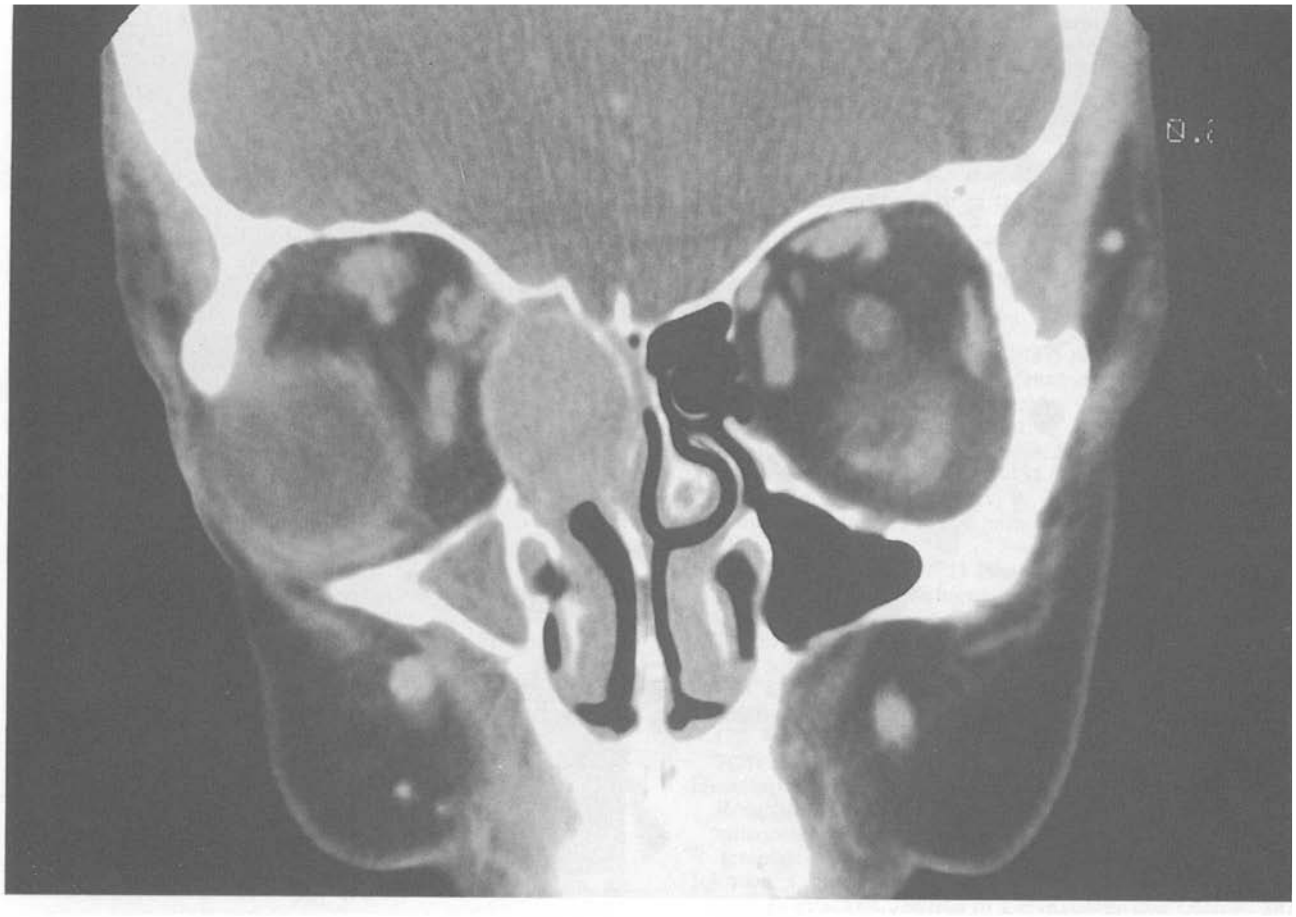

FIG. 2

Coronal CT scan showing enhanced mass occupying right ethmoid cells, extending to right frontal sinus and laterally into the orbit displacing globe inferiorly and laterally.

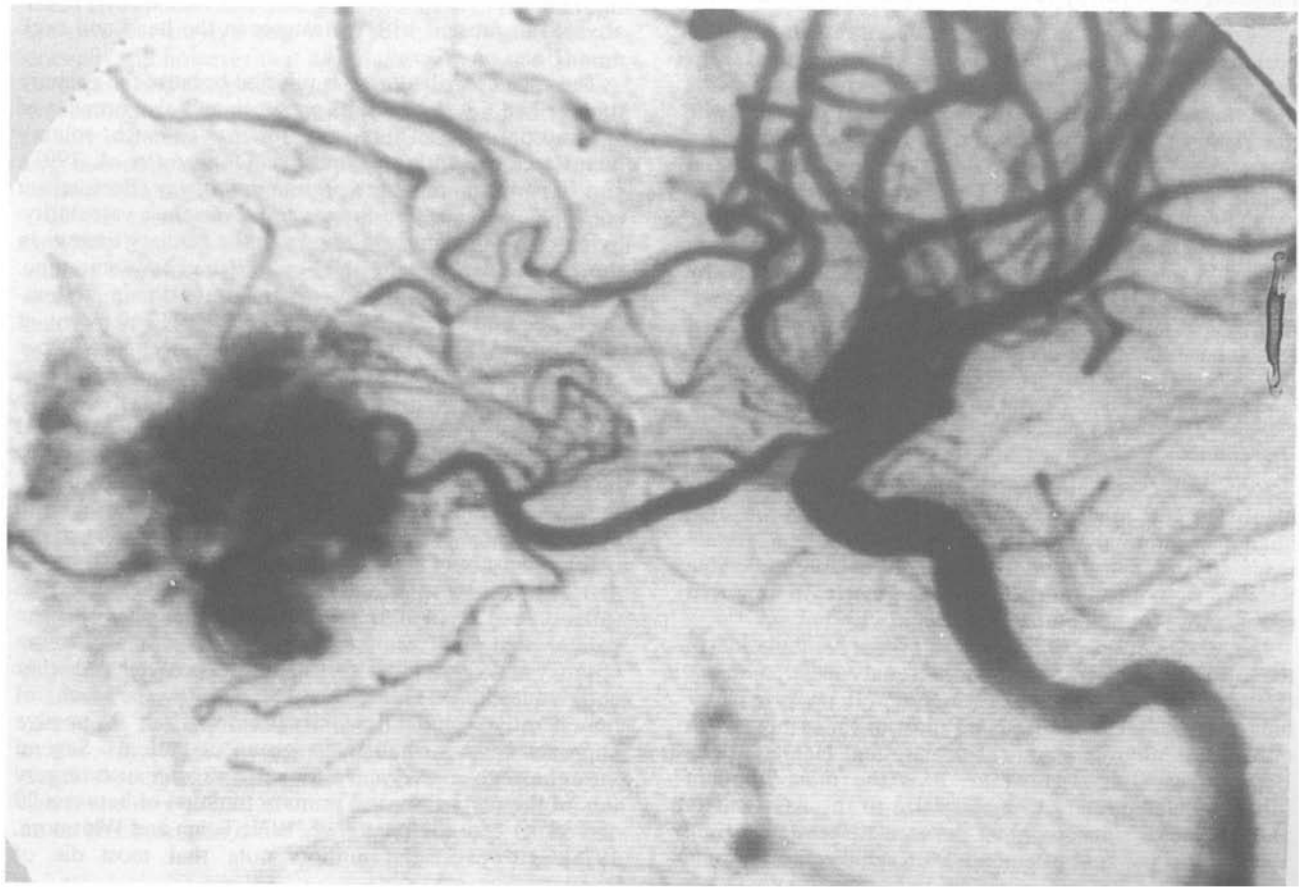

FIG. 3

Right internal carotid angiogram showing vascular nature of the metastasis and principal supply from right ophthalmic artery.

https://doi.org/10.1017/S0022215100131822 Published online by Cambridge University Press

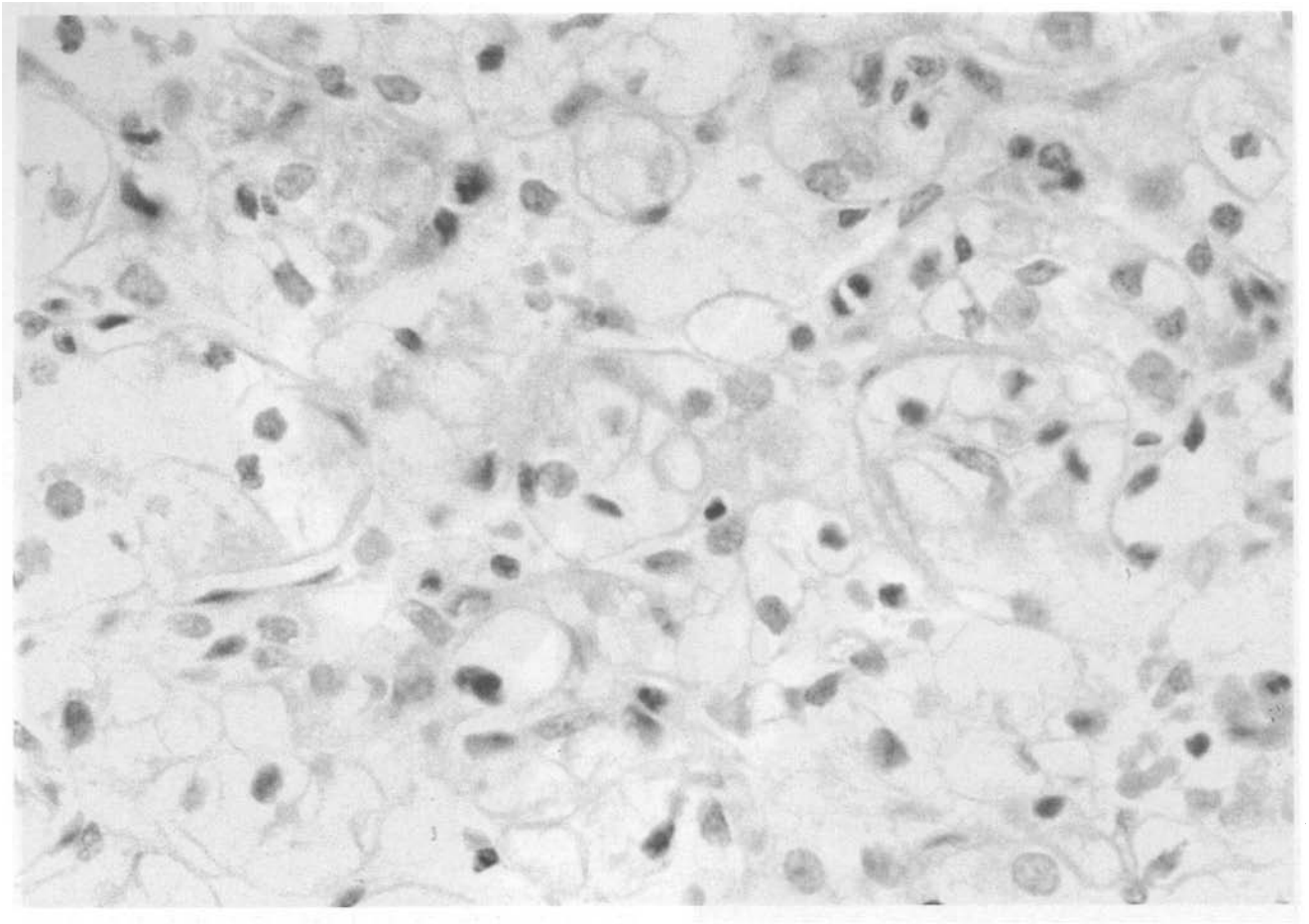

FIG. 4

Histology from the endoscopic biopsy of the metastasis showing renal cell carcinoma, with characteristically clear cytoplasm. (H&E; ×300).

Two weeks later, she had a craniofacial resection of her periorbital metastasis. This was done via a bicoronal scalp flap and craniotomy. The tumour was resected en bloc with dura which was adjacent to the metastasis. A mucocoele of the right frontal sinus was resected in addition to the metastasis. The supraorbital margin was reconstructed using split calvarial bone, both frontal sinuses were cranialized and the skull base defect was repaired with a pericranial flap. She was discharged eight days later. Since the lesion was invading bone, external beam radiotherapy (40 Gy) was administered in 15 fractions, although these lesions are not noted for their radiosensitivity. She had remained well 24 months post-operatively (Figure 5).

Discussion

Renal cell carcinoma characteristically metastasizes in a highly variable manner but it has a predilection for lung, bone and adrenal glands. The metastases are usually vascular and can themselves act in an unpredictable fashion, from being very locally aggressive to reported spontaneous regression (de Riese *et al.*, 1991).

Metastases from infraclavicular primary neoplasms to the head and neck are uncommon. Renal cell carcinoma is the third most frequent primary tumour (15 per cent) after lung and breast carcinomas, and most of these metastases occur in the thyroid gland (Miyamato and Helmus, 1973; Bassil *et al.*, 1985). However, it is the most frequent infraclavicular primary to metastasize to the nasal cavity and paranasal sinuses (40-50 per cent: Bernstein *et al.*, 1966). Eight per cent of patients with renal cell carcinoma are said to present with metastases in the head and neck area (Boles and Cerny, 1971).

The case described here is unusual because the primary tumour had not declared its presence and also because of its unusual site. There are reports of such solitary metastases presenting as epistaxis (Johnson *et al.*, 1993), and 70 per cent are said to present in this way (Bernstein *et al.*, 1966), which is not unexpected given their vascularity. Symptoms and signs resulting from the primary tumour in this disease can remain silent or obscured for some time. Indeed, the classical triad of costovertebral pain, abdominal mass and haematuria only occurs in 10 per cent of patients and, although haematuria does occur in 90 per cent, it is usually intermittent and microscopic (Robbins *et al.*, 1984).

It is generally agreed that an aggressive approach should be adopted in patients with a solitary metastasis from this tumour. Non-surgical treatment modalities such as chemotherapy, immunotherapy and radiotherapy have failed to improve survival significantly and, where possible, surgery is the mainstay of treatment (Dineen *et al.*, 1988). In the first reported case of surgery for a solitary metastasis (pulmonary) from a renal cell carcinoma in 1939, the patient died eight years later of ischaemic heart disease (Barney and Churchill, 1939). The success of this and other early anecdotal reports together with the lack of nonoperative modalities has promoted an aggressive approach to this small select group of patients. Several series have given five-year survival rates after such surgery (i.e. of the metastasis and primary tumour) of between 29 and 35 per cent (Skinner *et al.*, 1971; Tolan and Whitmore, 1975). However, the authors note that most die of

FIG. 5
Post-operative photograph of patient (at five months).

recurrent disease. A more recent series (Dineen *et al.*, 1988) gives poorer five-year survival rates (13 per cent) but the two-year survival rate is 41 per cent. The conclusion drawn is still however that aggressive surgery of a solitary metastasis and the primary does offer short-term survival and palliation. In the case of our patient, she clearly had a rapidly expanding mass of her periorbital margin causing her not only diplopia and pain but also considerable distress with regard to her facial appearance.

In the case described, a biopsy was obtained endoscopically. This is preferable as it avoids an external incision and the biopsy site can be resected en bloc with the tumour at the time of resection. The provisional diagnosis was of a mucocoele which is the commonest expansile soft tissue mass arising from the paranasal sinuses. There were some atypical features of this lesion, namely pain and some irregularity of the margins of the lesion on CT. Mucocoeles are usually associated with expansion and thinning of bone rather than focal bone destruction which is associated with malignant neoplasms. Adjacent soft tissue structures are displaced rather than eroded (e.g. periorbital fat will form a plane between a mucocoele that has expanded into the medial orbit and the medial rectus muscle). Mucocoeles are homogeneous, isodense with brain tissue and do not usually enhance unless infected (Hesselink *et al.*, 1979). Most primary malignant paranasal sinus neoplasms are not expansile in nature (Som and Shugar, 1980).

In this case, a mucocoele formed secondary to a renal metastasis. It is unusual for a paranasal tumour to present in this way (Hayes *et al.*, 1985). Having established the diagnosis, delineated the blood supply by angiography and also found no evidence of other metastases, we believe that an en bloc resection is the treatment of choice. It provides good palliation and also offers the best hope for long-term survival.

Acknowledgement

Mr Iain Robertson, consultant neurosurgeon, who jointly managed the care of this patient is thanked for his help.

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