CASE REPORT

Solitary calvarial haemangioma presenting as metastatic renal cell carcinoma

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SUMMARY

Renal cell carcinoma is the most common renal tumour in adult that often metastasises to the lung, liver or bone. Head and neck lesions are uncommon with no early warning signs and presents with overt metastases at primary presentation in 25%–30% of reported cases. The incidence of haemangiomas that suggest malignancy are similar to that of bone metastasis. Calvarial haemangiomas usually present as asymptomatic and discovered incidentally on imaging or postmortem examination. We report a case where an initial diagnosis of benign tumour of the skull was made based on clinical presentation and calvarial haemangioma on CT head but was confirmed as metastatic clear cell carcinoma of the kidney after histopathological results. Skull metastases are rare and present late in the course of the disease. It is unusual for metastatic lesion to be the primary presentation in a clinically silent renal cell carcinoma.

BACKGROUND

Renal cell carcinoma is the most common renal tumour in adult. It often metastasises to the lung, liver or bone. Head and neck lesions are relatively uncommon with reported cases of spread to the dorsal tongue, parotid glands, tonsils, skull, paranasal sinuses and thyroids.¹ There are normally no early warning signs and patients present with overt metastases at primary presentation in 25%–30% of the cases that have been reported.²

In a clinically asymptomatic patient with clinical and radiological investigation suggestive of a benign skull tumour, the diagnosis does not typically suggest a malignant tumour. There are multiple different diagnosis such as osteoma, osteoid osteoma, haemangioma and meningioma.³ The incidence of haemangiomas occurring in such a fashion is similar to that of bone metastasis, occurring as 3% each of all such incidental findings.^{4,5}

Calvarial haemangiomas usually present as asymptomatic and typically discovered incidentally on imaging or postmortem examination.^{6–9} Most patients seek medical attention when the mass is visible at about 1–2 cm.¹⁰ Common symptoms at presentation are due to compression of the mass on surrounding structures thus presenting with epidural haematoma, proptosis and cranial deformity.¹¹ Neurological deficit, such as dysarthria, drooling, facial neuropathic pain and new constant pressure-like headache,^{12,13} depends on the site of cavernous haemangioma.

Figure 1 CT head, transverse plane (A) and coronal plane (B) views demonstrating left-sided tumour with full thickness bone destruction. (C) Post-contrast CT head.

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Figure 2 MRI head demonstrating lesion with marked heterogeneous enhancement and some adjacent dural enhancement.

The presence of pulsatile tinnitus allows for multiple different vascular diagnoses. It can be classified by arterial, arteriovenous and venous cause. It is often caused by highly vascularised tumours of temporal bone (16%), venous of normal variants and anomalies (14%) and vascular stenosis (9%).¹⁴ This was the initial basis of our diagnosis of calvarial haemangioma in our patient who had clinically asymptomatic renal cell carcinoma.

CASE PRESENTATION

A 41-year-old man with no history of trauma presented with a 2-month history of pulsatile sensation on the left temple. The patient was otherwise fit and well with no other symptoms. On examination, there was a bruit present over the swelling but no focal neurology. An ultrasound was performed which detected a defect within the skull vault in the left frontotemporal region, within which there was a 3.1 cm highly vascular lesion. A CT scan of the head was performed for further assessment which detected a 40×27 mm avidly enhancing soft lesion centred on the lateral aspect of the left frontal bone. There was loss of the full thickness of the skull with small linear areas of calcification within the lesion (figure 1). A further MRI head scan was performed which demonstrated a mass that showed marked heterogeneous enhancement (figure 2). It was concluded that this may represent a calvarial haemangioma, and an urgent neurosurgical opinion was required.

INVESTIGATIONS

This case was referred to the Central Nervous System Multidisciplinary Team (CNS MDT) where a local diagnosis of calvarial haemangioma was agreed.

A subsequent excisional biopsy was undertaken (figure 3).

Histopathology of the specimen showed nests delineated by reticulin fibres of large pleomorphic clear cells surrounded by rich anastomosing thin capillaries and many large dilated

vascular channels (figure 4). Tumour cells were positive for renal cell carcinoma antigen, MNF-116, CK7, CD10 and PAX-8 (nuclear) but negative for EMA, CK20, S-100, CD31, CD34 and CD68. The diagnosis was confirmed metastatic clear cell carcinoma from kidney.

A CT of thorax, abdomen and pelvis was undertaken and demonstrated a 4.4 cm tumour arising from the left upper pole consistent with renal cell carcinoma. There was no locoregional lymphadenopathy and no focal liver or adrenal lesion (figure 5).

Figure 3 (A) Skin and temporalis dissected off tumour which eroded through the bone. (B) Gutter of bone excised around tumour which remains attached to dura below. (C) Underlying brain pia intact. Margin of dura excised around tumour to achieve a gross total resection.

Figure 4 (A) and (B). H&E stain. The tumour is composed of large nests and trabeculae of round and oval polygonal cells that have abundant clear or granular cytoplasm and small round and oval dark nuclei. The tumour has delicate branching vasculature and shows cystic and solid areas. The histological features are consistent with metastatic renal clear cell carcinoma.

DIFFERENTIAL DIAGNOSIS

Calvarial haemangiomas: osseous haemangioma, meningioma, dermoid/epidermoid cyst, multiple myeloma, brain metastasis.

TREATMENT

After confirmation of diagnosis, patient underwent left laproscopic radical nephrectomy and adrenalectomy

OUTCOME AND FOLLOW-UP

After surgery, MRI brain showed no evidence of recurrent or residual tumour in frontotemporal region.

Surveillance CT abdomen and thorax showed multifocal subcentimetre metastasis in anterior chest. Five millimetre left posas muscle lesion, two small lung nodules.

Patient was referred for consideration of high-dose IL-2. Patient is now commenced on high-dose IL-2.

DISCUSSION

There are a few cases of metastasis of renal cell carcinoma presenting as benign tumours. A case report by Koutnouyan *et al* found that their patient initially presented as epidermal inclusion

Figure 5 CT of thorax, abdomen and pelvis demonstrating tumour in upper pole of left kidney.

cyst on the forehead.¹⁵ A comprehensive review of literatures by Porter *et al* found that there were a number of cases of patient initially presenting as cutaneous metastases of renal cell carcinoma although solitary cutaneous manifestations of renal cell carcinoma are relatively uncommon. The available literatures emphasise the importance of looking for a primary renal cancer in the case of a cutaneous secondary clear cell adenocarcinoma.¹⁶

Skull metastases are rare and normally presents late in the course of the disease where primary renal carcinoma has already been identified initially. It is unusual for metastatic lesion to be the primary presentation in a clinically silent renal cell carcinoma. However, Jindal *et al* reported a case of calvari metastasis as a primary presentation of renal cell carcinoma.¹⁷

A retrospective study by Win looked at 20 patients with a medical history of cancer, who had solitary benign parietal lesions on CT head. The results showed that three patients had a skull lesion with a medical history of renal cell carcinoma of which only one was a haemangioma. This study concluded that benign parietal lesions can exist in patients with histories of cancer and that radiological features from CT scan can overlap between benign and malignant bone conditions and makes distinguishing between the two difficult. Hence, biopsy should always be undertaken when malignancy is suspected.¹⁸

Learning points

- ▶ Unusual presentation of a metastatic renal cell carcinoma.
- ▶ Preoperative radiology was misleading suggesting a benign tumour.
- ▶ Importance of histopathological studies in definitive diagnosis.
- ▶ Differential diagnosis of pulsatile tinnitus.

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