CASE REPORT

Renal cell carcinoma presenting as a cutaneous horn and nodules on the gingiva and scalp

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SUMMARY

A 63-year-old man presented with a pulsatile cutaneous horn on the nose and multiple angiomatous nodules on the gingiva and scalp, which appeared over 2 months. He had severe hypercalcaemia, lytic lesions in multiple bones and acute kidney injury. Excision biopsy from the gingival nodule showed a clear cell neoplasm. The bone marrow showed atypical cells with similar morphology. Imaging showed a 7 cm×7.5 cm mass at the upper pole of the left kidney with metastases to the bones, liver and lung. Immunohistochemistry was consistent with metastatic renal cell carcinoma. Renal cell carcinoma presenting as a cutaneous horn is extremely rare and to the best of our knowledge only one other case was found in the literature. There was visible regression in the size of the cutaneous horn and nodules following initiation of pazopanib therapy. However, he succumbed to his illness a month later.

BACKGROUND

Renal cell carcinoma presenting as head and neck skin nodules is rare. To our knowledge, there has been only one case report of a metastatic renal cell carcinoma presenting as a cutaneous horn, years following resection of the primary tumour and no report of such a primary presentation. In our patient, the cutaneous horn was the primary presentation of the malignancy along with other head and neck nodules. This case highlights the need to consider cutaneous metastasis of silent internal malignancies based on the accompanying clinical features. It also highlights the rare possibility of head and neck cutaneous nodules as well as cutaneous horn as the primary presentation of renal cell carcinoma.

CASE PRESENTATION

A 63-year-old man presented to our centre with multiple painless reddish nodules on the gums, and the scalp and a conical brownish crusted growth over the ala of the nose, all of which had appeared over 2 months. The nodules on the gingiva were 3 in number, reddish in colour and intermittently bled profusely when he brushed his teeth. The lesion on the nose was a brown, conical lesion with a reddish base, which had also bled spontaneously multiple times over the last 2 months. When he presented to our casualty, he was mildly disoriented with no history of headache, fever, seizures or neurological deficits. He had been taking calcium channel blockers for hypertension diagnosed a year ago.

The patient was moderately dehydrated at admission.

On examination, a hyperkeratotic, exophytic, pulsatile cutaneous horn measuring 2×1 cm with bright angiomatous base arising on the right ala nasi was noted (figure 1). Several angiomatous, firm, non-pulsatile nodules ranging in size from 1×2 cm to 4×3 cm were seen, scattered on the gingiva (figure 2), buccal mucosa, frontal and temporal scalp and right retroauricular area (figure 3). Examination of the abdomen did not reveal any hepatosplenomegaly or masses.

INVESTIGATIONS

The patient presented with severe hypercalcaemia (15.72 mg/dL) with normal phosphorous, low parathormone levels (<1.5 pg/mL) and an elevated alkaline phosphatase. He also had acute kidney injury and multiple lytic lesions on radiographs of the skull and long bones (figure 4). Initial workup for probable multiple myeloma revealed a normal serum electrophoresis pattern and absent urine Bence Jones proteins. Bone marrow trephine biopsy showed clusters of atypical cells suggestive of a metastatic carcinoma (figure 5). Histopathological examination of the excision biopsy from the lesion on the gingiva showed a neoplasm with clear cell morphology (figure 6). Non-contrast (due to acute kidney injury (AKI)) CT thorax showed multiple parenchymal and subpleural nodules in both lungs, lytic lesions in multiple vertebrae, the right scapula, ribs and sternum. It also showed a 7 cm×7.5 cm mass in the upper pole of the left kidney and multiple hypodense lesions in the liver. Ultrasound imaging of the abdomen confirmed the presence of a well-defined heterogeneously hypoechoic lesion with subtle vascularity at the upper pole of the left kidney. Immunohistochemistry of the gingival lesion was positive for vimentin, cytokeratin, PAX8 and CD10 (patchy positive) and negative for CK5/6 and S100. The patient was diagnosed with stage IV renal cell carcinoma.

DIFFERENTIAL DIAGNOSIS

Multiple myeloma was considered as a likely diagnosis on initial evaluation due to the hypercalcaemia, lytic lesions and renal failure. For the angiomatous nodules, we considered the possibilities of pyogenic granuloma, amelanotic melanoma or a metastatic malignancy arising from the lung or gastrointestinal (GI) tract.

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Figure 1 Cutaneous horn on the right ala nasi anterior and lateral view.

The severe hypercalcaemia was initially managed with hydration and loop diuretics. Steroids and calcitonin were added following the biopsy as the hypercalcaemia was refractory to the above measures. Following this, his calcium levels returned to normal and acute kidney injury showed an improving trend. His sensorium became normal with hydration and treatment of hypercalcaemia. Once the diagnosis of metastatic renal cell carcinoma was established, he was started on therapy with the multitargeted tyrosine kinase inhibitor pazopanib. The patient was offered palliative radiotherapy for the painful lytic lesions of the left femur; however, he did not wish to undergo radiation. During the hospital stay, there was visible regression in

Figure 2 Angiomatous gingival nodules.

Figure 3 Angiomatous scalp nodules.

the size of the nodules on the gingiva and the lesion on the nose.

OUTCOME AND FOLLOW-UP

Owing to stage of the disease, multiple metastases at presentation and rapid progression, poor prognosis was explained to the patient and his relatives in an appropriate setting. The patient wished to return to his hometown to continue therapy. He was discharged from our centre to continue pazopanib therapy and was advised to be on regular follow-up with an oncologist at his hometown. We learnt over telephonic correspondence that he had passed away a month later.

DISCUSSION

Renal cell carcinoma(RCC) accounts for 2.4% of all malignancies worldwide, with 337 860 new cases diagnosed in 2012.¹ In the European Union, 84 000 new cases of RCC and 35 000 attributable deaths occurred in 2012.² The incidence of RCC in Asian countries, particularly the Indian population, is lower than developed countries; however, data are limited.³ The reason

Figure 4 Lytic lesions of the skull and femur.

Figure 5 Histopathology of the bone marrow trephine biopsy showing atypical cell clusters.

for this could be that more tumours are picked up incidentally during abdominal imaging done for other reasons in developed countries. Data from the Surveillance, Epidemiology, and End Results (SEER) registry showed that at presentation 65% of cases were confined to primary site, 16% had regional spread, 16% had distant metastases and 3% were unstaged.⁴ 74.1% of patients diagnosed with RCC were alive at 5 years. In a study of 11 157 patients with metastatic RCC, the mean age at diagnosis was 64 years. Sixty-one per cent of patients had metastases to one site. The sites of metastases were to the lung (45.2%), bone (29.5%), lymph nodes (21.8%), liver (20.3%), adrenal (8.9%) and brain (8.1%).⁵ RCC is classified into distinct subtypes of which clear cell adenocarcinoma makes up 70% of all tumours. The other subtypes are papillary, chromophobe, oncocytic and collecting duct tumours.⁶ Cutaneous metastases occur in 4.6% of patients with RCC.⁷ Metastases to the head and neck region are rare and such metastases being the primary presenting symptom are even rarer. RCC is the third the most common malignancy below the clavicle to metastasise to the head and neck, following lung and breast.⁸ In a review of case reports of rare metastatic sites of RCC, among 12 cases of metastases to the nose and paranasal sinuses, 3 were at first presentation.⁹ In another review of tongue metastasis of RCC, among 29 cases described in the literature, only 4 were at primary presentations.¹⁰ To our knowledge, there has been only one case report, in 1983, of renal cell

Figure 6 Histopathology of the gingival lesion.

carcinoma presenting as a cutaneous horn. In that instance, the patient had undergone left radical nephrectomy for renal-cell carcinoma diagnosed 7 years ago.¹¹ In our patient, however, it was the primary presentation. The case report describes a hyperkeratotic lesion with an erythematous base which is very similar to the horn of our patient. The presence of an erythematous base, location on the nose and the rapid development of the lesion along with the presence of other nodules and severe hypercalcaemia indicated a malignant aetiology for the cutaneous horn. Other specific clinical features of a cutaneous horn that could indicate a possible malignancy are a low height-to-base ratio, pain, indurated base and lack of terrace formation that indicates rapid unorganised growth. Cutaneous horns associated with malignancy are common with squamous cell carcinoma in the vast majority and basal cell carcinoma in a few.¹² The angiomatous scalp lesions were very similar in appearance to a case reported in 2013.¹³ The findings of hypercalcaemia, lytic bone lesions and renal failure initially suggested a diagnosis of multiple myeloma. However, histopathological examination confirmed the final diagnosis. Cutaneous metastases of internal malignancy occur in 0.6%–10.4% of all patients with cancer and only represent 2% of all skin tumours.¹⁴ Cutaneous metastasis can be the primary presentation of a silent internal malignancy as in our patient. A high index of suspicion based on the clinical progression of the disease needs to be maintained in order to avoid delay in establishing a diagnosis. Histopathological examination is paramount in suspected cases, and along with immunohistochemistry provides valuable information directing further investigations. This case also highlights rapid progression of the disease with severe hypercalcaemia, AKI and extensive metastatic disease at presentation.

Learning points

- ▶ Renal cell carcinoma metastasising to the head and neck is rare and can present as pulsatile cutaneous horns or angiomatous nodules.
- ▶ A cutaneous horn can itself be a squamous cell carcinoma or rarely metastatic carcinoma.
- ▶ Cutaneous metastasis of internal malignancy needs to be considered in suspicious lesions with rapid clinical progression or other clinical and laboratory warning signs, warranting a careful search for possible underlying neoplasm.
- ▶ Histopathological diagnosis and immunohistochemistry is essential in such cases which can help in focused investigations to detect the primary neoplasm.

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