Extremely Rare Divergent Differentiation in Sarcomatoid Variant of Renal Cell Carcinoma Developing Lip Metastasis after Radical Nephrectomy

Manjeet Kumar*, Kalpesh Mahesh Parmar*, Santosh kumar1, Ritambra Nada4 and Abhishek Thakur5
1 Assistant Professor, Department of Urology, IGMC, Shimla, HP, India
2 Assistant Professor, Department of Urology, PGIMER Chandigarh, India
3 Additional Professor, Department of Urology, PGIMER Chandigarh, India
4 Professor, Department of Pathology, PGIMER, Chandigarh, India
5 Senior resident, PGIMER, Chandigarh, India

Abstract

Introduction: Renal cell carcinoma presents in 25% of patients with metastasis. The skin metastases are infrequent and are usually detected on follow up rather than with initial presentation. Only a few cases of lip metastasis have been reported in literature till date. We report an extremely rare case of renal cell carcinoma with divergent histopathology presenting as lip metastasis.

Case summary: A 75 years old male underwent Radical nephrectomy for Renal mass (T4N0M0). Histopathology was suggestive of a divergent Sarcomatoid variant of renal cell carcinoma. After 3 months of surgery, the patient presented with painless, hard swelling in the upper lip. Excision biopsy confirmed a divergent Sarcomatoid variant of renal cell carcinoma. The patient was started on tyrosine kinase inhibitors however patient succumbed to his aggressive disease after 6 months.

Discussion: Sarcomatoid with divergent pathology of renal cell carcinoma is rare. Renal cell carcinoma Sarcomatoid variant has aggressive metastatic behaviour. The skin metastases account for 1–3% of all metastases in RCC. The most common sites of metastases to skin reported are scalp and face. The outcome of patients with divergent pathology is dismal.

Conclusion: Metastasis to lip is a rare occurrence presenting in advanced carcinoma. Despite aggressive treatment, the prognosis is poor in divergent pathology of RCC.

Keywords: RCC, Lip metastasis, Divergent, Nephrectomy, Sarcomatoid.

Introduction

Renal cell carcinoma present in 25% of patients with metastasis. The skin metastases are infrequent and are usually detected on follow up rather than with initial presentation. Renal cell carcinoma commonly metastasizes to the lungs, liver, bone, adrenal glands, and the brain [1]. Only a few case report of metastasis to lip is present. We present a case of renal cell carcinoma with extremely divergent differentiation with lip metastasis.

Case summary

A 75 years’s man presented with swelling in the upper lip. The swelling was hard, located on the upper lip at the angle of the mouth. (Figure 1) The patient was following up case of right renal carcinoma (T2bN0M0) for which radical nephrectomy was done. At the time of surgery renal mass was localized and no metastases were seen on CECT chest and abdomen (Figure 1-3). Histopathology of renal mass was extensive divergent histopathology with a Sarcomatoid variant of renal cell carcinoma. The detailed histopathology included predominantly Sarcomatoid histopathology, some areas of squamous differentiation, and few foci of osteosarcoma with osteoid producing cells (Figure 4).

The patient was examined and investigated. The excision biopsy from lip was done and CECT abdomen was done. Biopsy of swelling was suggestive of a Divergent Sarcomatoid variant of renal cell carcinoma. CECT abdomen was suggestive of recurrence in the right renal fossa and multiple pulmonary nodules. In view of advanced
disease, the prognosis was explained and the patient was started on immunotherapy. The patient started on Pazopanib however the disease progressed, and the patient died within 6 months of diagnosis.

**Discussion**

Renal cell carcinoma especially Sarcomatoid variant has a tendency to metastasize early. Renal cell carcinoma has four major subtypes, clear cell carcinoma, papillary cell carcinoma, chromophobe cell carcinoma, collecting cell carcinoma. The most common metastasis from RCC is to lymph nodes, lungs, bones, brain. About 25 % of patients of RCC present with metastasis and another 25-50% will develop metastasis after nephrectomy. Skin metastases account for 1–3% of all metastases in RCC. About one fourth of patients in RCC present with metastasis and another one third develop recurrence after surgery [2, 3]. The most common sites of metastases to skin reported are scalp and face [4]. About 30 cases to face have been reported with the renal cell as primary, however, metastasis to lip reported in one patient. RCC metastases have been seen in the nose, lip, and hard palate, tongue and maxillary sinus [5]. Also, Sarcomatoid with divergent pathology of renal cell carcinoma is infrequently reported [3]. The management of these patients depends upon the systemic burden of disease. The facial lesion should be differentiated from basal cell carcinoma, hemangioma, pyogenic granuloma, etc. CT scan chest, abdomen, and brain or PET CT scan are done to identify metastasis. Excision biopsy is the gold standard to establish the diagnosis. The skin lesion almost always associated with metastases in other organs like lungs, bone, etc. The facial lesions should be excised however other lesions may be conservatively managed. If this is isolated metastasis aggressive resection is done followed by treatment with systemic therapy [6].

In a review of literature in Japan of 75 cases, the trunk (40%) was the commonest site for skin metastases, followed by scalp (25%),
and only 8% patients had metastases to face. In all these patients 24% had cutaneous metastases at the time of diagnosis [7].

Another case report describes a 55-year-old male with a single metastasis on the chin detected in follow up after surgery, the patient refused for further treatment and died 6 months later [8].

Other case highlights 55-year-old male presented with multiple metastases most notable were bleeding metastasis on the tip of the nose, liver, and lung. The patient died due to haemorrhage in brain metastasis on follow up [9]. Other cases mentioned in the literature include 61-year-old male with metastasis to ear. The patient was also having liver metastases at the time of diagnosis of RCC and developed recurrence of the skin lesions postoperatively after surgery [10].

Our case of lip metastasis was having metastases in the lungs, bone, and abdomen. The facial lesion was excised, and the patient was started on Tyrosine kinase inhibitor Pazopanib, however, the patient succumbed to the disease in view of extensive metastasis and variant histopathology.

**Divergent histopathology**

renal cell carcinoma with divergent differentiation. A) The photograph shows partially demarcated highly cellular renal tumour with adjoining spared renal parenchyma. B) The tumour predominantly displays Sarcomatoid areas in fascicles with foci of necrosis. C) Most of the areas exhibit osteosarcoma-like differentiation with osteoid producing tumour cells. D) At places, rhabdoid morphology in tumour cells is also evident. E) Small foci of squamoid differentiation are seen. F) Tumour is seen reaching the liver parenchyma indicating capsular breach and extension beyond Gerota's fascia. G) Vimentin immunostain performed shows diffuse strong cytoplasmic positivity in tumour cells. H) Pan-cytokeratin immunostain is negative in tumour areas; however, the periphery shows entrapped normal tubular component of renal parenchyma. I) CD10 immunostain is diffusely positive (cytoplasmic) in tumour cells (figure 5).

**References**


