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Case Study

Forty-three-year-old woman with mediastinal PEComa treated with chemoradiation therapy

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Abstract

The rarity of PEComa tumours has precluded any clinical trials, but surgery remains the most commonly reported treatment modality with just a few reports on treatment plans involving chemoradiation. We describe a patient with a mediastinal PEComa who presented with symptoms concerning for superior vena cava syndrome. She was deemed inoperable and was thus treated exclusively with chemoradiation therapy. The use of chemoradiation in the treatment of PEComa tumours is reviewed.

Keywords: chemoradiation; mediastinal tumour; PEComa; superior vena cava syndrome

INTRODUCTION

Perivascular epithelioid cells (PEC) are found in several distinct tumour entities described in the literature, such as angiomyolipoma, clear-cell sugar tumour, lymphangioleiomyomatosis and clear-cell myomelanocytic tumour, as well as in other sites.[1] Tumours with PEC predominance have been reported in a variety of anatomical sites including the genitourinary tract, gastrointestinal tract, lungs, kidneys and mediastinum.[2–4] PECs have a unique immunohistochemical (IHC) profile, staining with variable intensity for CD117, HMB45, HMSA-1, MelanA/Mart1, MitF, actin and desmin.[5] The mixed melanocytic/muscular IHC profile and the varied locations of PEComa tumours have often made rendering of an accurate diagnosis a challenge.[2]

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Surgical intervention is the most widely reported treatment; however, no clinical trials have been conducted to date, comparing this approach with radiation or chemotherapy. As not all patients are suitable candidates for surgery, and there are few case reports on the use of radiotherapy in the treatment of PEComas, a report on the use of radiotherapy as a primary intervention expands the scope of reported treatment approaches.

CASE REPORT

A 43-year-old Caucasian woman with a 20 pack-year history of tobacco was observed after she presented to her primary care physician with left arm swelling, pain and increased venous prominence concerning for superior vena cava syndrome. In addition, she complained of fatigue, acutely worsening shortness of breath after walking two blocks, intermittent headaches and light-headedness. Pertinent negatives on

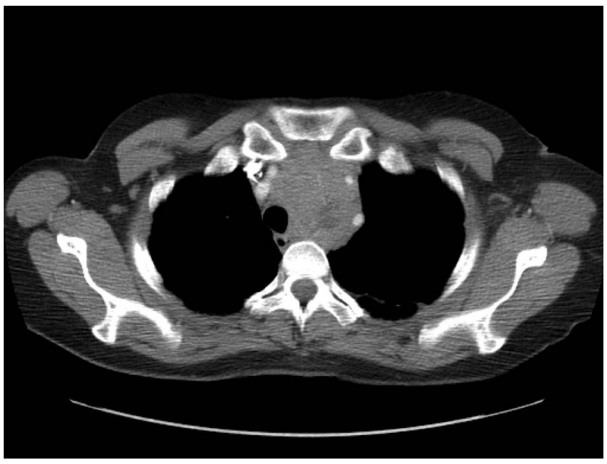

Figure 1. Patient's initial axial computed tomography with contrast of PEComa tumour located within the mediastinum.

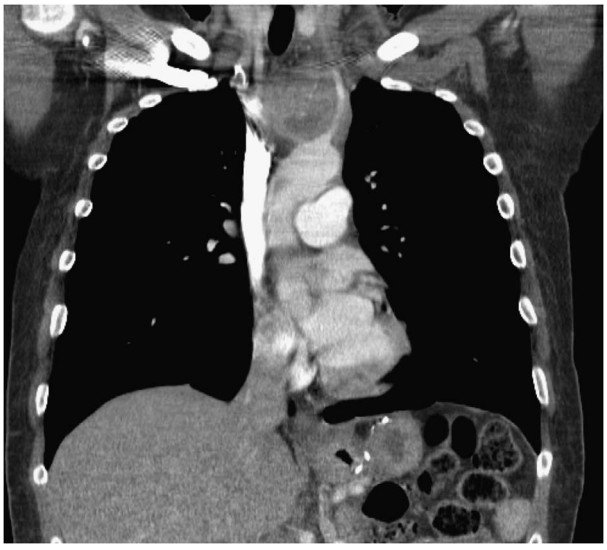

Figure 2. Coronal view of PEComa located within the mediastinum before treatment. Involvement of critical structures made the patient a poor candidate for surgical resection of the tumour.

review of systems included absence of cough, weight changes, fever or night sweats. Her medical history was unremarkable, and there was no known history of malignancy in her family. Her surgical history consisted of gastric bypass performed 6 years ago. Vitals were stable with oxygen saturation of 92%, and aside from mild left upper extremity oedema and superficial venous collateralization over her left upper chest, the physical examination was unremarkable.

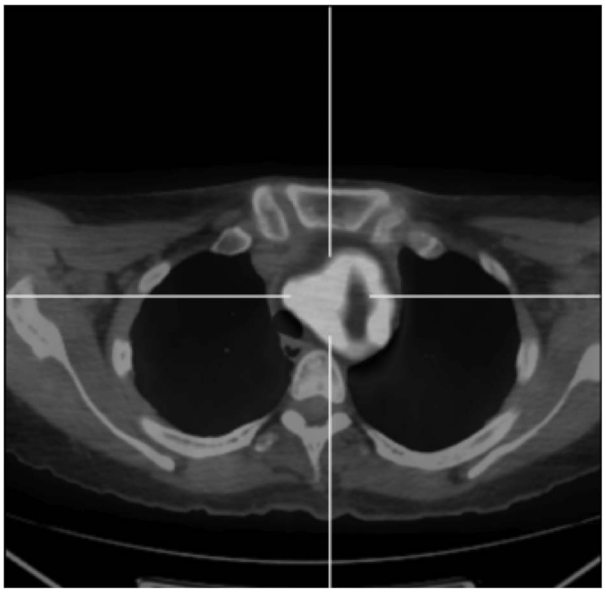

Figure 3. Axial staging computed tomography/positron emission tomography fusion demonstrating increased 18F-FDG avidity of the tumour.

Computed tomography (CT) revealed a $6{\cdot}8 \times 6{\cdot}4 \times 5{\cdot}8$ cm anterior mediastinal mass encasing the common carotid artery, with mass effect on the trachea and left brachiocephalic vein. There was no evidence of metastatic disease in the chest, abdomen or pelvis. She underwent endobronchial ultrasound-guided transbronchial needle aspiration of the mass, resulting in a preliminary diagnosis of non-small cell lung cancer (NSCLC) based on initial morphological features. Magnetic resonance imaging of the head showed no evidence of metastatic disease, and she was staged preliminarily at Stage IIIA NSCLC (T4N0M0).

Evaluation by cardiothoracic surgery found her to be a poor candidate because of the mass's involvement in the adjacent mediastinal structures. Owing to her acutely worsening shortness of breath and tracheal deviation, the decision was made to treat her urgently with palliative concurrent chemoradiation therapy with 61·2 Gy in 34 fractions with cisplatin and etoposide.

After three fractions of radiation totalling 5·3 Gy, initial IHC results returned positive for HMB-45 and MART-1, and negative for: PAN-Keratin, Mucin, CK 7, CK 20, Napsin, TTF-1

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and S-100. This profile was suggestive of melanoma.[7] An outside pathologist at a cancer hospital further corroborated the diagnosis. Positron emission tomography carried out at this time revealed abnormal uptake in the area of the mediastinal mass with a standardised uptake value of 11, which was consistent with melanoma.

The treatment plan was then modified accordingly. Daily treatments were increased to 2·5 Gy/fraction, with the goal of achieving a biologically equivalent dose (BED) of 100 Gy based on Phase III data from RTOG 83-05.[6] She received 9 Gy at 1·8 Gy/fx daily followed by 42·5 Gy at 2·5 Gy/fx daily for a total dose of 51·5 Gy, with a BED of ~100·5 Gy for the combined regimen. At this point she had not yet started on cisplatin/etoposide, and her chemotherapy was switched to temozolomide for radiosensitization, as well as for the high likelihood of metastatic disease.[7]

Additional, IHC staining results showed focally positive desmin and weak CD117, altering the diagnosis to PEComa.[1] By this time she had completed chemoradiation therapy, which she tolerated well. Swelling in her left upper extremity remained stable throughout the treatment and subsequent follow-up. Chest CT performed 5 weeks after completion of the therapy showed that the lesion had decreased in size to 6·1 × 5·8 cm, although there was an increase in prominence of a number of pulmonary nodules. The most recent restaging CT of the abdomen and pelvis showed no evidence of metastatic disease.

DISCUSSION

The standard of treatment for PEComas has not been well established, although it has been typically treated with surgical resection. There have been two case reports on pelvic PEComas being treated with surgery with adjuvant chemoradiation therapy. The rationales for post-operative radiation were not given.

In the first case, the initial clinical diagnosis was uterine leiomyoma, but a subsequent IHC profile was consistent with PEComa. The patient went on to receive post-surgical adjuvant chemotherapy followed by consolidation radiotherapy of an unknown dose. Unfortunately, there were bone metastases and local recurrence at 10 months' follow-up. She was reportedly in poor health at the time of the report's publication.[8]

The second patient presented with abdominal pain and vaginal spotting and was found to have a uterine PEComa, diagnosed via IHC profile. She eventually received an unspecified neoadjuvant chemotherapy regimen and surgical resection. This was then followed by 45 Gy of concurrent radiotherapy initiated during the third cycle of adjuvant vincristine, ifosfamide and doxorubicin chemotherapy. The patient was tumour free at 1·5 years.[9]

For our patient, the location and mass effect of the tumour were considered strong indicators for emergent treatment, although the final diagnosis was dependent on pending IHC stains. The location and involvement of critical anatomical structures precluded surgery, with chemoradiation being the only alternative. Suboptimal surgical conditions are likely to arise with PEComas, given their reportedly varied locations, and thus it is paramount that alternatives to surgical intervention be considered. As such, the local control achieved with radiation and temozolomide thus far is encouraging. Our patient is currently being considered for everolimus, a small molecule inhibitor of mammalian target of rapamycin, which has shown some promise in the treatment of some PEComas.[10]

Acknowledgements

None.

Conflicts of Interest

None.

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