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Extending Survival in a Patient with Extensive Stage of Small Cell Lung Cancer Presenting with Superior Vena Cava Syndrome as an Initial Symptom: A Case Study

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Abstract

Introduction: Superior vena cava (SVC) syndrome which is defined as the obstruction of blood flow through the SVC vein, often affecting patients with malignancy especially small cell lung cancer (SCLC) and is potentially an emergent condition needing prompt management. It is an ominous sign with poor prognosis and a mean of 8-10 months survival rate.

Case presentation: Here we present a middle-aged man who presented with fascial swelling and dyspnea with final diagnosis of advanced SCLC. Combination of multiple chemotherapy regimen, prophylactic brain radiation with administration of anticoagulant drugs were performed for him leading toward extension of his survival to 28 months.

Conclusion: These therapeutic approaches may lead to extending the survival in patients with SVC syndrome and SCLC.

Key words: Single-Case Studies as Topic; Small Cell Lung Carcinoma; Superior Vena Cava Syndrome; Survival

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INTRODUCTION

Superior vena cava (SVC) syndrome is defined as the obstruction of blood flow through the SVC vein (1). It is a kind of oncology emergency needing prompt management (2). Its symptoms such as fascial erythema, swelling and edema are secondary to upstream increase in venous pressure resulting from SVC vein occlusion leading to venous drainage impairment in the head and neck (3). It can potentially propel to cerebral edema and intracranial hypertension or airway compromise secondary to laryngeal edema (4). SVC syndrom usually affects patients with malignancy especially those with small cell lung cancer (SCLC) due to its central location and associated lymphadenopathy causing pressure effect over SVC vein (5). Although the underlying disease is somewhat sensitive to chemotherapy and radiotherapy, full recovery is often difficult to achieve. Here we present a middle-aged man who presented with SVC syndrome with final diagnosis of advanced SCLC who underwent several therapeutic methods leading toward extension of his survival to 28 months.

CASE PRESENTATION

A 54-year-old man presented with facial swelling, erythema and dyspnea lasting for 4 weeks. He also had productive cough with occasional bloody sputum. His past medical history was positive for 25-year cigarette smoking (50 packs a year). On physical exam he was a man with body mass index (BMI) of 24 and blood pressure of 115/75 mmHg, heart rate of 85 beats per minute, respiratory rate of 19 per minute and O2 saturation of 90% on room air. Facial plethora, neck swelling, purple discoloration of the face and neck with prominent external jugular veins and some collateral vessels on his face, neck and upper thoracic area were found (Figure 1). Other parts of physical examinations were unremarkable. Chest X-ray was requested which showed lobulated mediastinal widening and “S-golden sign” in right lung representative of concomitant right hilar mass and upper lobe atelectasis. Elevation of the right hemi-diaphragm was also seen with “juxtaphrenic peak sign” secondary to upper lobe atelectasis (Figure 2).

Contrast enhance computed tomography (CT) scan of the chest was performed which revealed a large mass about in right side of upper mediastinum extending to the right hilum. It causes significant narrowing and encasement of

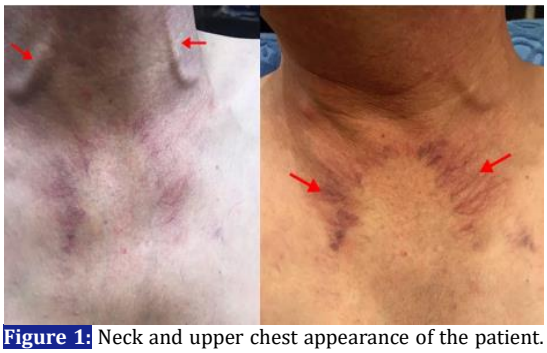

Figure 1: Neck and upper chest appearance of the patient. Engorgement and dilation of the superficial veins and extensive venous collateral circulation in the chest wall with also bilateral external jugular vein prominence and purple discoloration of neck and upper thorax is seen (red arrows)

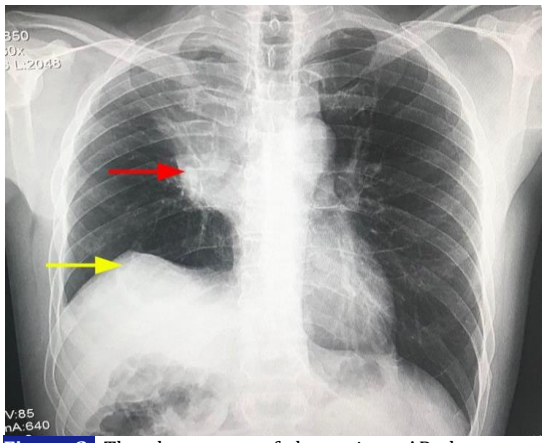

Figure 2: The chest x-ray of the patient. AP chest x-ray shows lobulated mediastinal widening and "S-golden sign" in right lung representative of concomitant right hilar mass and upper lobe atelectasis. (red arrow) elevation of right hemi-diaphragm is also seen (yellow arrow) with "juxtaphrenic peak sign" secondary to upper lobe atelectasis.

the SVC vein and right pulmonary artery. Elevation of the right hemi diaphragm was demonstrated with linear atelectasis at basal segments of the right lung representative of phrenic nerve invasion by the mentioned large mass (Figure 3). In visualized part of abdomen, there was enhancing heterogeneous mass involving the left adrenal gland suggestive of metastasis. Based on imaging findings advanced stage of lung cancer with metastasis (T4N2M1) was diagnosed.

Trucut biopsy was done under guide of CT scan. The final histopathologic result confirmed SCLC. Chemotherapeutic regimen was prescribed including 14 cycle platinum doublets protocol, Cisplatin and Etoposide. SVC syndrome associated symptoms improved significantly after the third chemotherapy session.

Ten sessions of the whole brain radiotherapy were done prophylactically. The patient had no neurological symptoms at time of brain radiation and subsequent brain magnetic resonance imaging (MRI) revealed no intracranial metastasis. Whole body positron emission tomography scan (PET-scan) was done to determine the response to treatment after the 8th chemotherapy. It showed a central 18 F-FDG avidly uptake mass in right upper lobe representative of primary SCLC (SUV max=8.3). Right para-tracheal 18 F-FDG-avidly uptake metastatic adenopathy was seen (SUV max=6.6). Another FDG avid uptake mass within the right adrenal gland was also note (SUV=6.2) (Figure 4).

According to PET-scan no new metastasis was occurred and partial response to treatment was achieved. SVC syndrome symptoms reappeared after cessation of chemotherapy regimen. Chemotherapeutic regimen was changed to Ifosfamide – Etoposide therapeutic protocol. Intravenous anticoagulant medication was also prescribed. The patient's syndrome responded well with new chemotherapeutic protocol. After that patient presented with bilateral pleural

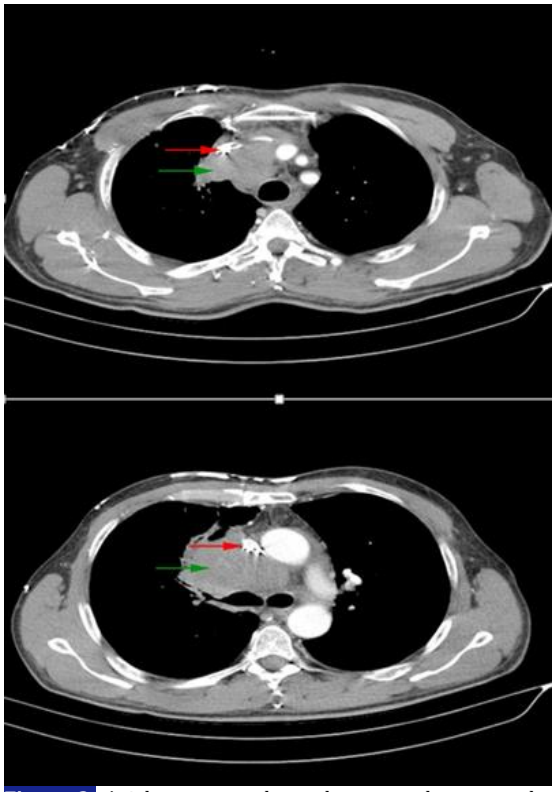

Figure 3: Axial contrast-enhanced computed tomography scan (CT scan) of upper mediastinum revealed large mass (green arrows) with pressure effect and encasement over compression of the SVC (red arrows) and extending to right hilum.

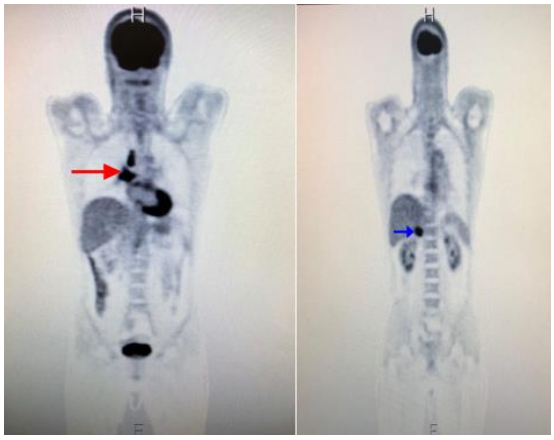

Figure 4: Frontal view of positron emission tomography scan (PET-Scan) showed an avidly uptake 18F-FDG mass lesion in the right upper lobe and the right para-tracheal that are primary lung mass and its metastatic para-tracheal lymph node respectively (red arrow) and another mass in the right adrenal gland (blue arrow)

effusion which pleurodesis with bleomycin was performed. After two years, the patient presented with neurologic symptoms including seizure and slurred speech. Subsequent brain MRI found multiple intracranial metastasis. Unfortunately, despite all therapeutic modalities, the patient was expired almost 28 months after initial presentation.

Discussion

SVC syndrome usually presents in patients with underlying malignancy, leads to an ominous prognosis; but it sometimes responds to chemotherapy with or without radiotherapy (6, 7). CT-scan is a valuable modality for detection of possible underlying malignancy in patients presenting with SVC syndrome (8).

SCLC accounts for 20-25% of all lung cancer cases. Due to its central location and associated lymphadenopathy, causing pressure effect over SVC vein, it can cause SVC syndrome (5). It is an aggressive form of lung cancer with tendency toward dissemination and early metastasis (9). Limited-stage of SCLC is cured by combination of chemotherapy with radiation therapy; in about 20% of the patient's cure can be achievable in early stages. The mean survival rate for patients with limited-stage of the disease is nearly 18 months (10). SCLC cases presenting at advanced stage of the disease are primarily treated with chemotherapy, with a high initial response rate of about 70% and a complete response rate of about 30% but with a median survival time of approximately 10 months (11, 12).

Nowadays platinum and etoposide combination is the accepted standard chemotherapeutic regimen (13). Thoracic radiotherapy (TRT) is another accepted therapeutic method for patients with limited stage of the disease (14). Adding TRT would improve the survival rate approximately 5% in comparison to chemotherapy alone. TRT performed simultaneously with chemotherapy is more effective than sequential therapy (15). Furthermore, the survival benefit is greater if TRT is given earlier in the course of chemotherapy (16). Patients with SCLC whose symptoms persist or progress after first-line chemotherapy have a high mortality rate. Second-line therapy may produce a modest clinical benefit. Few studies have revealed that there is usually partial response to the SCLC after the first line of chemotherapy; but, unfortunately, we see the tumor regression infrequently, and most cases usually need second line therapy (17, 18).

In a case report from the Netherland the authors reported a SCLC case with paraneoplastic nephrotic syndrome who had a 7-month survival rate. They present the same picture of our case (but they didn't start a prophylactic brain radiotherapy for the patient at all) and after 5 months of first line chemotherapy, the patient had a brain metastases and then expired after two months (19); this is in contrast to our results about the survival rate.

In another case from China, the authors presented a patient with transformed SCLC from lung adenocarcinoma with metastasis to the brain and then liver during the chemotherapy treatment cycles. They also did not do the prophylactic brain radiation in the first time that the diagnosis was confirmed, and finally they saw rapid progression of the tumor to other organs like brain and liver and the patient died within a short period of time (20).

Although the combination of SVC syndrome and SCLC leads to survival rate of less than 8 months, here we report a case in which combination of multiple chemotherapy regimen, prophylactic brain radiation with administration of anticoagulant drugs contributed to the improvement of survival to 28 months.

Conclusions

Multiple combination of chemotherapeutic regimen with radiation therapy, prophylactic brain radiotherapy, and anticoagulant administration can leads to extending the survival in patients with SVC syndrome and SCLC, and may provide bright horizons in the treatment of similar patients.

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AUTHORS' CONTRIBUTION
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CONFLICT OF INTEREST
None declared

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