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# A Rare Cancer with a Rare Occurrence: Pulmonary Adenocarcinoma with Enteric Differentiation as a Cause of SVC Syndrome

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## Presentation

Superior Vena Cava Syndrome, also known as SVC syndrome, is an uncommon occurrence resulting from reduced blood flow to the right atrium from SVC obstruction. Patients experience multiple nonspecific symptoms such as headache, facial redness/swelling and shortness of breath. This was first noted in 1757 by the Scottish anatomist and physician William Hunter, where he recognized syphilitic aortic aneurysm as a cause [1]. Subsequently in the mid-1900s, a review done on 274 cases of SVC syndrome suggested 40% of these cases were due to infections (tuberculous mediastinitis and syphilitic aneurysms) [2].

In more recent times, a significant majority (60-90%) can be attributed to lung cancers alone, however, we now see the rise of benign causes of this syndrome from intravascular devices and pacemakers (some studies reporting up to 40% of cases) [1,3-7]. Metastatic disease-causing SVC syndrome has been shown to be relatively infrequent, accounting for under 10% [8]. Here we show a patient who developed SVC syndrome from an extremely rare malignancy, pulmonary adenocarcinoma with enteric differentiation (PAED). PAED is a subtype of pulmonary adenocarcinoma in which the enteric component exceeds 50%. To differentiate the primary cancer in this subtype, immunohistochemistry with markers such as CK-7, CK-20, CDX-2 and TTF-1 are used. However, in those who are CK-7 negative, differentiation becomes difficult and clinicians must base their assessment on imaging/endoscopic findings. Here we present the 4th case in English literature of CK-7 negative PAED, but the first described case of PAED associated with SVC syndrome.

## Assessment

Patient was a 69-year-old male with a known history of stage IV prostate cancer diagnosed in 2012 that was treated with leuprolide. He was asymptomatic until 2018 where he was found to have an elevated PSA with new back pain. A CT scan showed multiple large lung masses, one of which appeared to be related to the pleura of the right lung. Bone scan was evident for multiple bone and soft tissue metastases. Given the rare propensity of prostate cancer to metastasize to the lungs, he was referred for a CT guided lung biopsy.

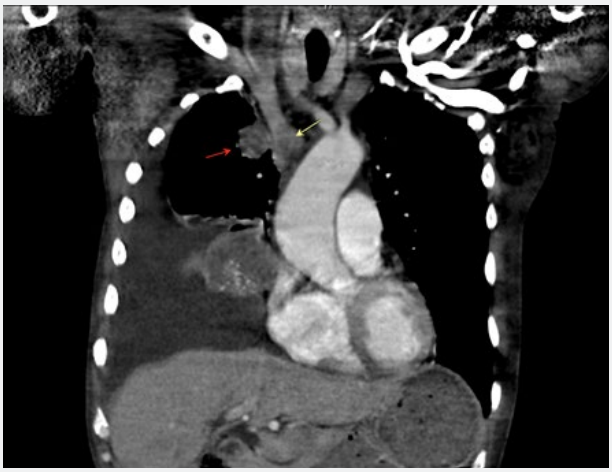

**Figure 1**: CT scan of the chest with IV contrast showing a mass (red arrow) obstructing the SVC and a distal thrombus (yellow arrow)

Prior to the biopsy the patient presented to the ED with dizziness and facial swelling for two weeks. He was hypotensive, tachycardic, and tachypneic with a large right pleural effusion on chest x ray. CT scan of the chest showed a lung mass partially compressing the superior vena cava at the area of its junction with the innominate vein with evidence of thrombosis distally (Figure 1). Therapeutic thoracentesis was performed alleviating the patient's symptoms.

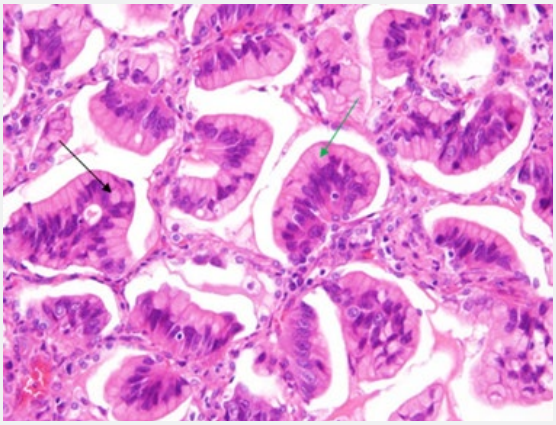

**Figure 2**: Histologic section of CT guided lung biopsy of right upper lobe mass. Green arrow shows glandular epithelial cells indicative of adenocarcinoma. Black arrow shows goblet cells indicative of enteric differentiation. >50% of biopsy shows goblet cells - a key feature of PAED.

CT guided lung biopsy was then done, and results were supportive of enteric/intestinal-type adenocarcinoma (cytokeratin 7 -, cytokeratin 20 +, and CDX 2 +) (Figure 2 will be pathology of lung mass). Patient at this time had no known history or suspicion of an enteric malignancy.

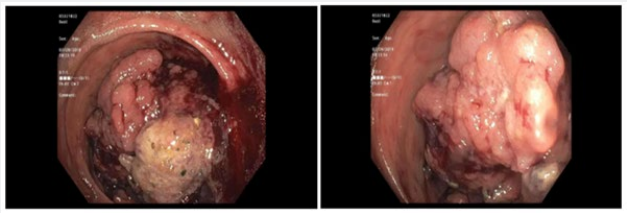

**Figure 3**: Colonoscopy images showing a large fungating mass completely obstructing the entire rectal lumen.

His recovery course was then complicated by hematochezia. Colonoscopy was evident for a large fungating mass that was obstructing the entire rectal lumen (Figure 3). Biopsy showed a tubular adenoma with high grade dysplasia. Colo-Rectal Surgery then performed a transverse loop colostomy to relieve the obstruction.

## Management

The primary origin of metastatic adenocarcinomas can be difficult to diagnose. The use of immunostaining over the past several years has greatly increased our ability to make an accurate diagnosis. Staining with antibodies to CDX-2, cytokeratin 7, cytokeratin 20, and thyroid transcription factor (TTF-1) have all proven to be useful [1]. These help to differentiate between primary lung cancer, metastatic lung cancer and colorectal adenocarcinomas.

CDX-2 is a protein found in transcription factors on intestinal epithelial cells. Cytokeratin 20 (CK-20) is a major cellular protein found in mature enterocytes and Cytokeratin 7 (CK-7) is a protein found in glandular epithelial cells of the breast and lungs. This patient's biopsy stained CK 20 +, CDX2 + and CK 7 -. This is indicative of an enteric malignancy.

The patient experienced a rather complicated disease course. Initially it was presumed he had a recurrence of his prostate adenocarcinoma with metastases. However, prior to his lung biopsy, he presented with SVC syndrome. Later the same admission, he developed hematochezia where a colonoscopy showed a large fungating mass obstructing the entire rectal lumen. The biopsy and immunohistochemical stains (of the lung and rectal mass) were consistent with rectal adenocarcinoma. He underwent a diverting loop colostomy, radiation to his rectal mass and was placed on chemotherapy with bevacizumab/irinotecan. Bevacizumab, an angiogenesis inhibitor, and irinotecan, an inhibitor of DNA replication, is a widely used chemotherapy regimen in metastatic colorectal cancer.

At the time of diagnosis, around 50% of patients with colo-rectal malignancies will have metastases or non-operable disease [9]. Those with unresectable disease, such as this patient, have a 5% or less 5-year survival rate [10]. In metastatic colo-rectal cancer, combination chemotherapy is now the standard of care in those with good functional status (ECOG 0-1). Overall survival rates in these patients can be up to 30 months (median) [11].

## Authorship

All authors had access to manuscript and a role in writing it.

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