

A Case Of Wegener's Granulomatosis With An Unusual Initial Presentation Of A Pancoast Syndrome

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Abstract

Non-small cell lung cancer is the most common etiology of Pancoast syndrome (Pancoast's syndrome). There is only one other similar case of Wegener's granulomatosis presenting initially as a Pancoast tumor reported in the medical literature. "Classic" Wegener's granulomatosis is a form of systemic vasculitis (polyangiitis) that primarily involves the upper and lower respiratory tracts and the kidneys. A "limited" form, with clinical findings isolated to the upper respiratory tract or the lungs, occurs in approximately one-fourth of cases. Up to one-third of patients with Wegener's granulomatosis who present with pulmonary involvement may not have respiratory symptoms. Other organ systems that may become involved in addition to the kidney include arthralgias, conjunctivitis, episcleritis, uveitis and vesicular, purpuric, and hemorrhagic lesions. Nervous system manifestations include mononeuritis multiplex, cranial nerve abnormalities, external ophthalmoplegia, tinnitus, hearing loss. This is a unique presentation and it highlights the importance of cytologic, histologic, and microbiologic examination of tissue that can be attained by percutaneous biopsy of the Pancoast tumor causing the neurologic symptoms.

INTRODUCTION

Pancoast syndrome [1] is a clinical constellation of an apical lung lesion and neurologic sequelae caused by compression of the adjacent lower trunk of the brachial plexus and sympathetic trunk in the superior pulmonary sulcus.

Non-small cell lung cancer is the most common etiology of Pancoast syndrome (Pancoast's syndrome). Other causes include nonepithelial neoplasms, infections, lymphomatoid granulomatosis, vascular aneurysms, amyloid nodules, and cervical rib syndrome. Because of the wide variety of diseases that can produce Pancoast syndrome, a histologic diagnosis is mandatory before initiating definitive treatment. There is only one other reported similar case reported in the medical literature [2].

CASE REPORT

60 yr old Caucasian female with Past Medical History of dyslipidemia presented with Left shoulder pain described as constant, dull in nature, radiating down her left arm, not associated with motion. She denies any history of trauma. No fever, chills, redness or swelling or weight loss. She had a smoking history. She denied any alcohol or illicit drug use. She worked as a clinical psychologist and she was happily married. She admitted to having a chronic cough with intermittent arthralgias and morning stiffness. Her physical

exam was unremarkable. Her complete blood count and comprehensive metabolic profile was unremarkable and revealed no evidence of any anemia or renal disease. Urinalysis revealed hematuria but no casts or proteinuria.

Radiographs done revealed evidence of multiple pulmonary nodular densities in the Left superior pulmonary sulcus, the left lower lobe and the right upper lobe. CT Scan [Figure 1] confirmed the presence of a 9.5 × 8.3-cm right middle lobe mass, 3.7 × 2.9-cm and 3.1 × 2.2-cm right lower lobe masses, a 3.6 × 2.0-cm left upper lobe mass, and many diffuse smaller nodules throughout the lung fields.

Pathology of CT guided biopsy specimens revealed chronic granulomatous inflammation and no neoplastic process. ESR was 73 and C-ANCA was positive. Other collagen vascular diseases blood tests like Rheumatoid factor levels, Anti-Nuclear Antibody titers, P-ANCA, anti-myeloperoxidase and protease-3 abs were negative.

Diagnosis of Wegener's granulomatosis was made and the patient was started on corticosteroids and oral cyclophosphamide 1 mg/kg/day which resulted in resolution of pulmonary nodules. Subsequently patient had resolution of her previously persistent left shoulder pain.

Figure 1



DISCUSSION

Pancoast syndrome is an entity that is most commonly associated with bronchogenic carcinomas of the lung apex, most frequently squamous cell carcinoma, followed by adenocarcinoma and large cell carcinoma, and only rarely small cell lung carcinoma. Because of their peripheral location, the initial presentation of these tumors is dominated by features referable to invasion of local structures, principally nerves and the body wall.

Shoulder pain is the most common presenting symptom, related to the involvement of the lower trunk of the brachial plexus, the first 3 ribs, the vertebrae, or the parietal pleura. Horner's syndrome, recurrent laryngeal and phrenic nerve involvement are less common features. Pulmonary manifestations, such as cough, hemoptysis, and dyspnea, occur less often than with centrally located endobronchial lesions. [3]

Nonneoplastic conditions include neurogenic thoracic outlet syndromes and inflammatory conditions (eg, Wegener granulomatosis and amyloidosis).

Wegener's Granulomatosis presenting as Pancoast syndrome has only once been previously reported in the literature.

WEGENER'S GRANULOMATOSIS

"Classic" Wegener's granulomatosis is a form of systemic vasculitis (polyangiitis) that primarily involves the upper and lower respiratory tracts and the kidneys. A "limited" form, with clinical findings isolated to the upper respiratory tract or the lungs, occurs in approximately one-fourth of cases.

Up to one-third of patients with Wegener's granulomatosis who present with pulmonary involvement may not have

respiratory symptoms.

Other organ systems that may become involved in addition to the kidney include [4]:

Joints (myalgias, arthralgias, arthritis)

Eyes (conjunctivitis, episcleritis, uveitis)

Skin (vesicular, purpuric, and hemorrhagic lesions)

Nervous system (mononeuritis multiplex, cranial nerve abnormalities, external ophthalmoplegia, tinnitus, hearing loss)

Heart (pericarditis, myocarditis, conduction system abnormalities)

Less commonly, the gastrointestinal tract, subglottis or trachea, lower genitourinary tract (including the prostate or ureter), parotid glands, thyroid, liver, or breast

Nonspecific complaints of fever, anorexia, weight loss, and malaise may accompany upper or lower airway disease. In addition, signs and symptoms related to involvement of other organ systems include ocular inflammation, nasal congestion, joint tenderness and or effusion, and rash. Skin lesions include palpable purpura, ulcers, vesicles, papules, and subcutaneous nodules.

Lab findings are generally nonspecific in Wegener's granulomatosis including those with pulmonary involvement. Common abnormalities include leukocytosis, thrombocytosis ($>400,000/\text{mm}^3$), elevated ESR and anemia of chronic disease.

The urinalysis is normal in limited Wegener's granulomatosis. However, patients with renal involvement typically show an elevation in the BUN and plasma creatinine concentration, mild to moderate proteinuria, and RBC, WBC and cellular/ hyaline casts.

Most patients (90%) with Wegener's granulomatosis have C-ANCA, characterized by autoantibodies directed against serine proteinase 3. In some circumstances, a positive ANCA may provide the only clue to the correct diagnosis and may prompt decisive diagnostic and management decisions.

In addition to its diagnostic utility, the level of the C-ANCA has been used to follow the disease course.

DIAGNOSIS

Our case report highlights the management difficulty presented by multiple pulmonary masses in which a tissue diagnosis proves elusive. Diagnosis of pulmonary masses relies on cytologic, histologic, and microbiologic examination of tissue. This is vital because not only does it help in diagnosis but it also impacts on treatment. For apical tumors, percutaneous biopsy is reported as the modality of choice. [5]

CONCLUSION

Although the initiation of treatment for apical lung lesions was once advocated without the acquisition of a precise histologic diagnosis, [6] the recognition that Pancoast syndrome can be caused by a wide variety of conditions mandates that an accurate diagnosis is made in any pulmonary mass that appears clinically and radiologically to

be a bronchogenic carcinoma. CT-guided percutaneous biopsy remains the best modality of choice, but where the diagnosis remains elusive and uncertain, open biopsy is required.

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