Case Report

Shoulder Pain in an Elderly Smoker May be an Alarm: a Rare Case of Pancoast Tumor

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Abstract:

Patients with shoulder pain commonly present to a general practitioner and/or in the orthopedic department. In some instances, pain in the shoulder region can indicate serious underlying pathology. This article describes an elderly smoker presenting with progressively worsening shoulder pain not conforming to musculoskeletal, neuropathic or mechanical type pain and with poor response to conventional pain management protocols. Evaluation with meticulous history, physical signs, chest imaging findings, and histopathology confirmed the diagnosis of a rare clinical condition the Pancoast tumor.

Keywords: Pancoast tumor, Pancoast syndrome, Horner's syndrome, apical lung mass, lung cancer

DOI: https://doi.org/10.47648/jswmc2021v1102-09

JSWMC 2021[11(02)] P: 72-77

Introduction:

The term Pancoast tumor or superior sulcus tumor of the lung defines a wide range of tumors invading the apical chest wall and producing a characteristic syndrome named 'Pancoast-syndrome'.[1] Histologically they are commonly non-small cell carcinomas and are rare, comprising 3-5% of all lung cancers. Smoking is the predominant risk factor, just as for other lung cancers.[2] It is more common in men, and the average age at examination is in the sixth decade of life.

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Because of its distinct anatomical location, Pancoast tumor may present with characteristic clinical features that result from its local invasion of the brachial plexus, stellate ganglion and other adjacent structures.[1,2,3] Shoulder pain has been reported as being the first presenting feature of a Pancoast tumor in 90% of cases. It is of interest that the original description of a Pancoast tumor in 1932 by Henry Pancoast and Tobias was of shoulder pain due to an apical mass.[4]Missing a diagnosis is common due to lack of clinical suspicion even in the present time.

Case Summary:

An 80-year-old, smoker, normotensive, non-diabetic farmer presented with worsening right shoulder pain radiating down to right arm and upper back for the last 6 months. The pain was of moderate to severe intensity, dull aching in nature, and not fully relieved by medication, rest or physiotherapy.He had a history of smoking, consuming 40 pack year. He complained of occasional cough over the past years which became productive in the last month. He gave no history of fever, breathlessness, hemoptysis, chest pain, joint pain, oro-genital ulcers or limb

weakness. He was afebrile and gave no suggestive history of tuberculosis contact but lost 5 kg of his body weight over the past few months. On examination, he was mildly anemic, had finger clubbing (Fig. 1), and had normal blood pressure (120/70) mmHg, pulse (80beats/min), and core body temperature (98°**F)**. Evaluation of the nervous system revealed, partial ptosis (Fig. 2), miosis and enophthalmos of the right eye, and right hemifacial anhydrosis with no other deficit. On examination of respiratory system, dull percussion notes, low intensity vesicular breath sound with a few coarse crepitations were found over the right upper chest. Examination of the musculoskeletal system did not show any sign of arthritis, deformity, restricted movement or muscle weakness in the right shoulder or elsewhere. There was no organomegaly, thyroid abnormality, skin rash or lymphadenopathy and the rest of his systems revealed normal findings.

His routine blood tests were normal except for low hemoglobin (11 gm/dl) and raised ESR (80mm in first hour). His X-ray right shoulder was unremarkable but chest radiograph showed hyper inflated lungs and right apical opacity(Fig. 3). Sputum culture yielded growth of pseudomonas species sensitive to Ciprofloxacin, Levofloxacin, Amikacin, Imipenem, Meropenem and Gentamicin. Computed tomographic (CT) scan of the chest revealed complex findings comprising of consolidation without air bronchogram in the right upper lung field with peripheral densities suggestive of broncho-alveolar infiltrates. Parenchymal opacities were extended from right paravertebral region to infra-hilar regions. CT guided FNAC from lung lesion showed cellular material composed of atypical epithelial cells arranged singly and in small clusters. Individual cells had large nuclei, prominent nucleoli with abundant cytoplasm and features were compatible with non-small cell carcinoma of the lung.The patient received treatment with Amikacin for his chest infection but declined any further evaluation and specific management of his cancer.

An apical malignant tumor of right lung, with ipsilateral Horner's syndrome (ptosis, miosis, enophthalmos, anhydrosis) made up the diagnosis of a rare clinical condition, the Pancoast syndrome due to Pancoast tumor of lung along with super-imposed pseudomonas pneumonia.

Fig 1: Clubbing

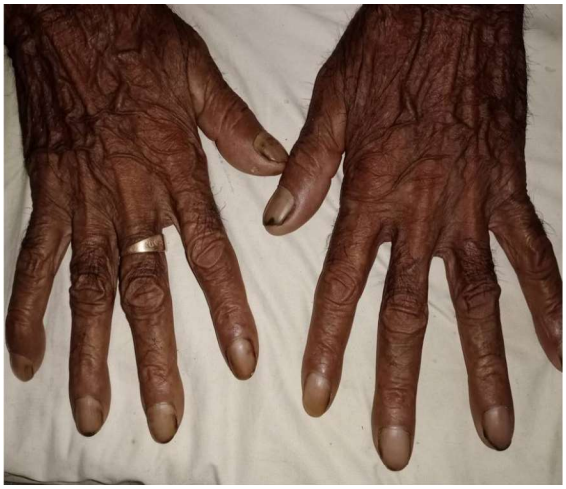


Fig 2: Ptosis

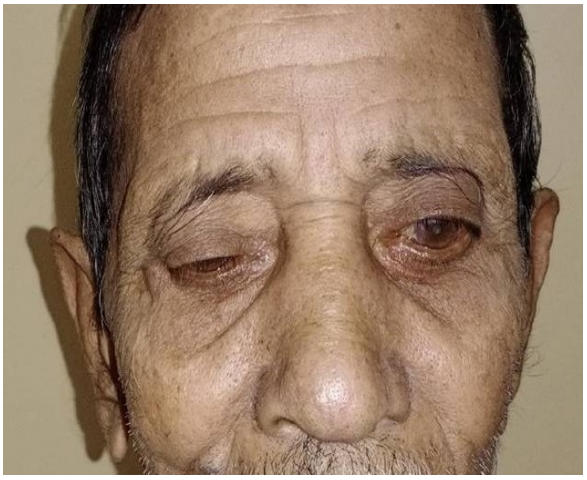


Fig 3: Chest X-ray showing right apical opacity (arrow)

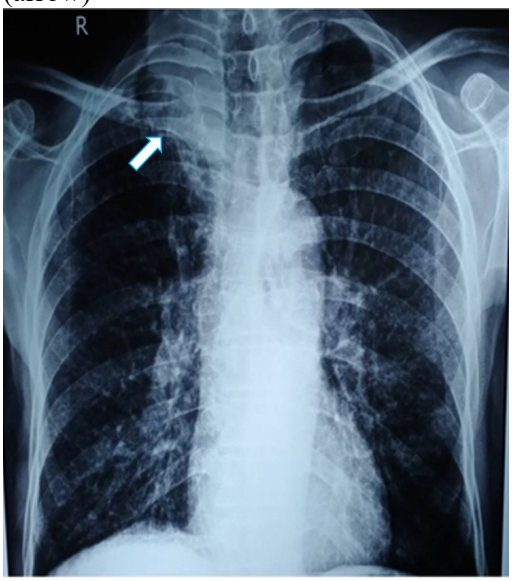
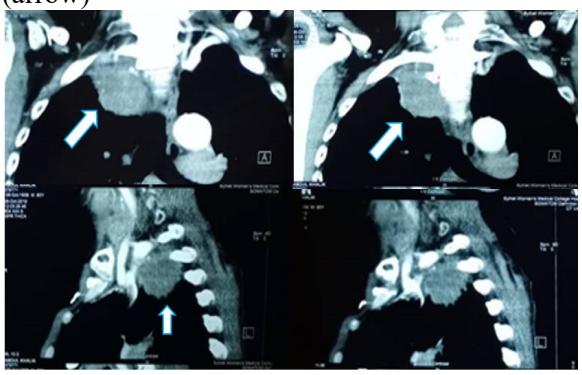


Fig 4: CT chest showing right apical mass (arrow)



Discussion:

Musculoskeletal pain is a common presenting complaint in medical practice and comprises one fifth of all musculoskeletal cases and shoulder pain is the commonest of all such pain.[8,9] And on rare occasions, a serious underlying etiology like Pancoast tumor (PT) is revealed.[10] PT is defined as a tumor of the lung that invades any structure of the upper part of the chest like the first rib, brachial plexus, sympathetic chain and stellate ganglion near the apex of the lung.[1,5]And, Pancoast syndrome(PS)is the constellation of signs and symptoms associated with tumors that involve the superior pulmonary sulcus and clinically consists of pain and tingling sensation of the shoulder and arm along the distribution of the ulnar nerve and ipsilateral Horner's syndrome (constriction of pupil, ptosis, enophthalmos and hemifacial anhydrosis).[6,7]Anatomically speaking, Horner's syndrome is usually pre ganglionic and is due to the interruption in the cervical sympathetic innervation to the eye.[6] The causes of Horner's syndrome can be threefold, based on the distribution of anhidrosis – i) anhidrosis affecting the face, arms and trunk indicating a central cause, ii) anhidrosis affecting the face only indicating a preganglionic cause and iii) absence of anhidrosis indicating a postganglionic cause.[6,7] Our patient had asymmetric anhidrosis only of the right side of his face and trunk or limbs were spared.

The most common initial symptom associated with PT is sharp posterior shoulder pain produced by invasion of the brachial plexus, parietal pleura, fascia, first and second ribs or vertebral bodies.[8] There may also be pain in the axilla or subscapular area on the affected side with radiation to the head-neck area, across the chest, and/or to areas down the medial aspect of the arm along the distribution of ulnar nerve.[4,5,7,9] As superior sulcus tumors do not involve the central tracheobronchial tree, signs-symptoms like cough, dyspnea, and hemoptysis that are commonly associated with non-small cell carcinoma of lung, are usually absent in PT.[10]

In our case, the patient had occasional productive cough that can be explained by concomitant presence of chronic obstructive pulmonary disease with bacterial infection evidenced by positive sputum culture. Most PTs are unilateral, but bilateral lesions have been reported in rare instances.[11] Events such as recurrent laryngeal nerve palsy[12], superior vena

cava syndrome[13], and airway obstruction[14]may occur late in the course of the disease. The principal symptoms of PT are the consequences of local invasion.[3,7] Though PS can be attributed to infection, it is unlikely in our patient because he had obvious proof of having a malignant lesion.[15,16]

The most common cause of PS is non-small cell lung cancer (NSCLC). Squamous cell carcinoma is the leading type, followed by adenocarcinoma and large cell carcinoma.[17,18] The Moffit Cancer Center Florida USA, reported that in 2012, of the examined 17,860 lung cancer patients, 550 cases had PT. Cases with PT were divided into squamous cell carcinoma(52%), adenocarcinomas (23%), large-cell carcinomas (20%) and only 5% small-cell carcinomas.[19] However lung cancers are not the only causes of PS. A wide range of other benign and malignant conditions may be responsible for its causation (Table 1).[19,20]

Table 1: Differential diagnosis of apical lung masses with Pancoast syndrome

Malignant causes:	**Primary bronchogenic carcinoma:** Non-small cell carcinoma **Non-bronchogenic primary lung cancer:** adenoid cystic carcinoma, hemangiopericytoma, mesothelioma **Metastatic carcinoma:** carcinoma larynx, cervix, urinary bladder, thyroid **Hematologic malignancy:** plasmacytoma, lymphoid granulomatosis, lymphoma, myeloma
Benign causes:	**Bacterial infections:** Staphylococcal and Pseudomonal pneumonia, Actinomycosis, Tuberculosis **Fungal infections:** Aspergillosis, Cryptococcus, Allescheriasis **Parasitic:** Echinococcus **Others:** Neurofibromatosis, cervical rib syndrome, pulmonary amyloidoma

Chest X-ray findings of PT may show unilateral apical cap of more than 5 mm thickness, asymmetry of both apical caps of more than 5 mm, an apical mass, and bone destruction. Typical and common finding in chest radiograph is, a thin 'plate' on the apex of the lung in the area of the superior sulcus.[1,3] Lordotic views and X-ray of the cervical and upper thoracic spine may provide a better determination of the tumor.[4] However, 1.7% of patients with PT, have a normal plain chest radiograph.[2]

Sputum cytology is positive in 10-20% patients with superior sulcus tumor.[1,8] As these tumors are peripherally located, the diagnostic yield from sputum cytology is less than those located more centrally. Fiber optic bronchoscopy is an important tool for collecting biopsy specimens from different lung lesions.[18] But the superior sulcus tumors are usually not visible through endoscopy because of their peripheral locations, and so, biopsy or brushings must be obtained under fluoroscopic guidance. Diagnostic yield with flexible fiber optic bronchoscopy is 40-60%. The most common diagnostic method for a PT is transthoracic needle aspiration. In one report of 27 cases of PS, a fine needle aspiration was positive for carcinoma in 26 out of 27 patients. In multiple studies, CT scan of chest was found to be 60% sensitive, 65% specific, and 63% accurate in determination of the local extent of tumor.[19]The clinical picture with chest radiograph findings of an apical lesion, frequently provides the presumptive diagnosis of a PT with a high degree of accuracy (>90%). However, confirmation by tissue diagnosis is always important.[10] The non-resolving shoulder pain and the presence of Horner's syndrome on clinical examination in our patient, guided us towards exploring more. And this is why we called for chest radiograph, CT and fine needle cytology which served with the diagnosis straight. So, this should be kept in mind that musculoskeletal pains may not always be as benign as it seems to be.

There is a 2-3% incidence rate of small cell carcinoma and occasional non-bronchogenic malignant lesions mimicking PT (Table 1). Fine needle aspiration has replaced, to a large extent, core needle biopsy and both have an accuracy rate of greater than 95%. Ultrasound-guided biopsies have the advantage of less radiation exposure and, real-time imagining, and is particularly beneficial in patients who cannot hold their breath for long.[1]

When a potential PT is suspected, some studies suggest that the diagnostic evaluation should begin with bronchoscopy. If adenopathy is present, a scalene lymph node biopsy and/or mediastinoscopy should be done.[11]These procedures have the dual roles of establishing a diagnosis and staging the mediastinal lymph nodes. In the absence of mediastinaladenopathy or when a tissue diagnosis by bronchoscopy is not possible, a radiology-guided needle biopsy will confirm the diagnosis in most of the cases.[9,10] Finally, if a definitive diagnosis is still evasive, thoracoscopic biopsy, open biopsy, supraclavicular biopsy, and/or formal surgical exploration may be required, depending on the extent of the tumor.[1] Superior sulcus tumor requires the same staging evaluation as other lung masses such as CT chest or MRI with contrast of the brachial plexus and thoracic inlet including the first two ribs.[7]Apart from confirming a diagnosis, histological samples are important for molecular profiling and treatment planning.[19]

We could not proceed with further evaluation and specific curative management steps due to disinclination of the patient. But theoretically, when recognized early, PT is treatable with chemo-radiotherapy followed by surgical resection.[10,11] With treatment, 2-year survival is around 55%-70%.[7] In the series from MD Anderson for 5-year survival prediction, worse prognostic factors were- (a) weight loss ≥5%, (b) vertebral body involvement, (c) supraclavicular involvement, and (d) advanced stage of disease.[8]Also, it is found that, when a malignant superior sulcus tumor causes PS manifested by presence of Horner's syndrome, a poor prognosis is implied. A 5-year survival is expected to be around 10% in patients with Horner's syndrome and around 30% in patients without it.[15] At least, with this piece of knowledge, a poor outcome is postulated for the case reported here.

Conclusion: Physicians should remain alert about missing a diagnosis of Pancoast tumor in patients with persistent shoulder and upper back pain with a background history of smoking. Moreover, a chest X-ray should be advised along with X-rays of shoulder and spine in all such patients.

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