Management of Lung Cancer Invading the Superior Sulcus

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KEYWORDS

• Superior sulcus • Pancoast • Non–small cell lung cancer • Apical • Tumor

KEY POINTS

- The first cases series of superior sulcus tumors was reported by Dr Pancoast, a radiologist, in the early twentieth century.
- For decades, superior sulcus tumors were thought to be distinct category of slow-growing lung cancers. They are now recognized, however, as non–small cell lung cancers that are distinct from nonapical tumors in their location only and not their biology.
- Modern treatment is centered around neoadjuvant chemoradiotherapy followed by surgical resection with the goal of achieving complete pathologic response and R0 resection.

HISTORY

> *There is an unusual but apparently infrequent type of intrathoracic growth occurring in the apical region, yet found with sufficient frequency in my experience to warrant a collective report of the cases encountered.*
>
> *—Henry K. Pancoast, MD*[1]

In 1924, Dr Pancoast described a series of patients with apical intrathoracic tumors in his landmark case series.[1] A radiologist by training, Dr Pancoast documented 5 instances of apical shadows on chest radiograph (CXR) that were diagnosed as intrathoracic tumors, either pathologically or clinically.[1] Despite the pathologic diagnosis of lung cancer in 1 of these tumors, Dr Pancoast concluded that these apical tumors were either pleural endotheliomas or sarcomas in origin.[1]

Dr Pancoast went on to become the first president of the American Board of Radiology.[2] In 1932, he gave a chairman's address at the annual session of the American Medical Association.[3] In his address, he described 7 total cases of "superior sulcus pulmonary tumors."[3] These superior sulcus tumors shared the following key characteristics: (1) an apical shadow along with posterior destruction of 1 or more of the first 3 ribs on CXR and (2) a distinct clinical presentation now commonly referred to as Pancoast syndrome.[3] This syndrome was described in his address as shoulder and arm pain in the C8-T2 distribution, wasting of the hand muscles, and Horner syndrome on the affected side.[3]

Unbeknownst to Dr Pancoast, one of his contemporaries, Dr Tobias, described the same clinical lesion and symptoms in 1932.[4] Unlike Dr Pancoast, Dr Tobias correctly identified the origin

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of these tumors as bronchogenic carcinomas.[4,5] In subsequent decades, the origin of these superior sulcus tumors was confirmed by other investigators as primary lung cancer.[6–10]

DEFINITION

All patients in Dr Pancoast's original case series presented with Pancoast syndrome.[1,3] Subsequent investigators also only included patients with superior sulcus tumors associated with classic symptoms in their case series reports.[10–13] As treatment paradigms began to change and surgical resection became an important component of multimodality therapy, investigators began to include any patients with apical lung cancers in their studies regardless of Pancoast syndrome, leading to inconsistent use of the term, *superior sulcus tumor*.[14–21] The lack of consistency led Detterbeck[8] to propose a formal definition of superior sulcus tumors in 2003. Detterbeck defined a superior sulcus tumor as a "lung cancer arising in the apex of the lung that involves structures of the apical chest wall," regardless of symptoms.[8] In his proposal, the apical chest wall was defined as the level of the first rib or higher. Chest wall structures included the parietal pleura, ribs, vertebral bodies, vessels, nerves, and nerve roots.[8] The American College of Chest Physicians has subsequently endorsed this definition as have other modern investigators.[5–7,22] The term, superior sulcus tumor, was rejected by many investigators in the decades that followed Dr Pancoast's landmark articles because the superior sulcus was not considered an embryologic or anatomic structure.[23] In modern literature, however, the terms, *Pancoast tumor*, *Pancoast-Tobias tumor*, and *superior sulcus tumor*, are now used interchangeably.[5–9,22]

CLINICAL PRESENTATION

It has been estimated that superior sulcus tumors represent less than 5% of patients with lung cancer.[22] One prospective database study identified superior sulcus tumors in 3.2% of patients with newly diagnosed lung cancer.[15] The incidence of lung cancer in the United States has been estimated at 228,190 cases per year.[24] Based on these numbers, it can be estimated that approximately 7300 patients present with superior sulcus tumors per year in the United States.

Almost all patients who present with a superior sulcus tumor are symptomatic. In 2 reported series that detailed the presenting symptoms of patients, the numbers of asymptomatic patients were only 5% and 1.5%, respectively.[14,15] The most common reported symptom is shoulder pain.[9,10,14,15,25] One study that looked at 139 patients who underwent surgical resection over 16 years at a single institution reported that 127 (91.4%) of patients presented with shoulder pain.[14] In another study, 107 of 131 patients (82%) with superior sulcus tumors presented with shoulder pain. As originally described by Dr Pancoast, shoulder pain commonly radiates to the arm, axilla, or scapula.[14] Different compartments may give rise to different presentations of arm pain. Pain in the anterior chest wall, shoulder and upper limb, and axillary distributions are typical of anterior, middle, and posterior compartment tumors, respectively.[26] Approximately 25% of patients present with Horner syndrome (24%–32% in the case series discussed previously[14]). Only approximately 10% of patients present with classic Pancoast syndrome (shoulder/arm pain, Horner syndrome, and hand muscle wasting).[10] Other less common symptoms include hemoptysis, weight loss, pneumonia, dysphonia, superior vena cava syndrome, and paraneoplastic syndromes.[5,9,10,14,15]

DIAGNOSIS

In his original case series, Dr Pancoast described the difficulty of identifying superior sulcus tumors on plain CXR, noting that a majority of tumors in his series were missed on initial interpretation of the CXR.[1] Although CXR remains important in incidental detection of or initial screening for superior sulcus tumors, any clinical suspicion for a superior sulcus tumor should be investigated with higher-resolution imaging studies. A reasonable first test to establish a diagnosis of superior sulcus tumor is a high-resolution, contrast-enhanced chest CT scan.[27] Chest CT not only can establish a diagnosis of superior sulcus tumor but also provide information on regional lymph node and thoracic metastases.[27] In addition, despite the multitude of imaging options available today, contrast-enhanced CT remains the best modality to assess tumor involvement of 2 specific structures. First, it provides superior visualization of bony invasion involving the ribs and thoracic spine.[27,28] Second, contrast-enhanced CT provides excellent resolution of the upper thoracic vascular structures that may be invaded by tumor.[27]

In the past several decades, the use of MRI in assessing superior sulcus tumors has become widespread.[27–29] MRI provides superior assessment of tumor extension into the surrounding soft tissues.[27–29] In particular, MRI is the best modality to detect tumor involvement with the brachial plexus nervous system as well as the vertebral foramina and spinal cord.[27] For this reason, it is currently the imaging modality of