Pancoast Tumors

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KEYWORDS

• Pancoast tumor • Superior sulcus tumor
• Brachial plexus • Hand function • Neurolysis

Pancoast tumors are bronchogenic cancers occurring at the lung apex (also known as the superior pulmonary sulcus), that produce symptoms and signs including hand, arm, shoulder and chest pain; hand weakness, numbness and wasting; and Horner's syndrome. The natural history, if left untreated, is rapid death within months of diagnosis.

The treatment of superior sulcus tumors has evolved over the past 170 years, and now includes chemotherapy, radiotherapy, and surgery, with modern treatment providing significant long-term survival for many patients. The standard surgical treatment of these tumors has hitherto involved resection of any anatomic structure(s) invaded by tumor, including the lower trunk of the brachial plexus, and/or the C8 and T1 nerve roots. This has resulted in significant disability with loss of hand function. We have adopted a different approach to the surgical management, with extensive neurolysis and preservation of all involved brachial plexus elements, with the intention of preserving hand function, without compromising patient survival.

HISTORICAL BACKGROUND

In 1838, Edward Selleck Hare, House-surgeon to the Stafford County General Infirmary in England, reported the first case of an apical lung tumor in a letter to the Editor of the Medical Gazette, in which he described a 40 year old man with

> *"pain, tingling and numbness along the course of the ulnar nerve of the left arm…. pain through the left shoulder…. the pupil of the left eye became contracted; and the levator palpebrae ceased to perform its office."* [1]

In addition to finding a palpable neck lump, Dr. Hare described the onset of neurologic decline related to spinal cord compression, and ultimately death, 3 months following the onset of symptoms. Description of the treatments used at the time included

> *"leeches and blisters were repeatedly applied over the tumor, but without any effect. When the extract of belladonna was applied the pupil of the left eye recovered its natural size for a time."*

After describing the post-mortem findings, including

> *"the tumor lay upon the brachial plexus, being firmly attached to the spine at the origin of the third and fourth nerves of the plexus, both which were inseparable from it,"*

Dr. Hare attempted to explain all of the clinical findings including:

> *"the connexion of the disease with the distress and paralysis along the course of the ulnar and median nerves is obvious, and is evidence in confirmation of the assertion that these nerves can be traced through the plexus to the last cervical and first dorsal nerve."*

However, he was unable to explain all.

> *"The paralysis of the levator palpebrae, which receives a branch from the third pair; the contraction of the pupil… cannot be referred to any direct communication between the structural disease and these several affections… and is most frequently displayed in persons*

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Neurosurg Clin N Am 19 (2008) 545–557
doi:10.1016/j.nec.2008.07.002
1042-3680/08/$ – see front matter

of a nervous temperament… and is parallel to what occurs in hysteric females, of which I have seen many examples.''

The excellent understanding of the brachial plexus, median and ulnar nerve anatomy is overshadowed by Hare's lack of knowledge of the sympathetic supply to the eye, and it is for this failure to understand the patient's physical signs that Hare is not credited with the eponymous title. However he never had the opportunity to pursue the sympathetic supply to the eye any further, because Hare died of typhus the day before his letter was published.[2]

In 1924 Henry K. Pancoast, a radiologist from Philadelphia, presented a paper at the seventy-fifth Annual Session of the American Medical Association in Chicago, and published his findings in which 3 cases of ''apical chest tumors'' were reported.[3] Dr. Pancoast stated that

''the neoplastic condition is unusual for the reason that it produces referred nerve phenomena in the upper extremity which may be very misleading to the clinician and roentgenologist in their search for the cause.''

Pancoast's subsequent, and more notable paper was his chairman's address, read before the section on Radiology at the American Medical Association meeting in New Orleans in 1932.[4] At that time, Pancoast had adopted the name ''superior pulmonary sulcus tumor.'' He reported seven cases with

''clinical and roentgenographic characteristics which would warrant the inclusion of the tumors in a group which could be recognised as a pathologic entity.''

Further description included

''the tumors in question seemed to occur at a definite location at the thoracic inlet, were characterized clinically by pain around the shoulder and down the arm, Horner's syndrome and atrophy of the muscles of the hand and presented roentgenographic evidences of a small, homogeneous shadow at the extreme apex, always more or less local rib destruction and often vertebral infiltration. Death occurred as a result of what seemed to be a comparatively trivial growth without detectable metastases roentgenographically.''

Pancoast stated that Horner's syndrome is ''an essential manifestation'' as it occurred in all seven of his reported cases, whereas hand weakness or wasting only occurred in three cases. He commented that

''pain on the ulnar aspect of the forearm suggested the supply of the internal cutaneous from the eighth cervical and first and second thoracic nerves. Muscular wasting involved the interosseous muscles and those of the hypothenar eminence and the web of the thumb. This would correspond to an ulnar supply from the eighth cervical and first thoracic nerves. Horner's syndrome would place the lesion in the region of the common trunks from the eighth cervical and first thoracic at least.''

Unlike Hare before him, Pancoast succeeded in correlating the clinical and radiological findings to describe this disease entity, however, he did err in his pathologic assessment. He believed that these tumors took ''origin in an embryonal epithelial rest,'' and ''one can practically rule out primary lung cancer.'' With regard to prognosis, Pancoast noted that these tumors are ''rapidly fatal'' with death occurring 11 to 14 months after the onset of symptoms. He believed that these tumors were not ''subject to surgical removal,'' with irradiation being the only possible treatment option.

Pancoast originally referred to these tumors as apical chest tumors,[3] but subsequently used the term superior pulmonary sulcus.[4] There has been confusion as to the correct nomenclature, and the exact definition of the superior pulmonary sulcus. The pulmonary sulcus is ''the costovertebral gutter whose superior limit is the first rib arch and whose inferior limit is the insertion of the diaphragm in the thoracic cage.''[5] The terms apical lung tumor, superior sulcus tumor, and Pancoast tumor may be used interchangeably.

In 1946 Peter Herbut and John Watson,[6] from Jefferson Medical College Hospital, Philadelphia, added their 17 cases of thoracic inlet tumors to the 134 published cases, although, of the 151 total cases, only 100 cases were carcinoma of the lung. Herbut and Watson noted that the average time from symptom onset to death was 10.5 months, and that ''Roentgen therapy so far appears to be of no avail,'' and ''to date the disease has been 100 per cent fatal.'' For a detailed historical review of the published cases of that era, the interested reader is referred to Herbut and Watson[6] for cases published unto 1946, and Chardack and MacCallum[7] for a review of published cases between 1946 and 1953. It was not until the 1956 publication from William Chardack and James MacCallum,[8] of Buffalo, New York, that surgical treatment became a viable option. They presented the first report of surgical resection with postoperative irradiation and a 5 year survival. The surgical technique was presented as ''a long