Cancer Association of South Africa (CANSA)

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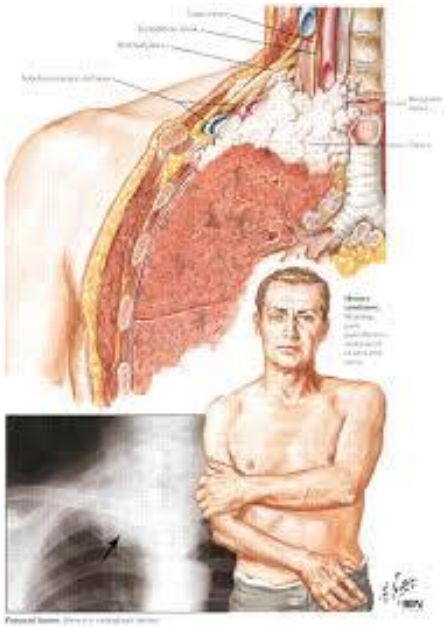
Fact Sheet on Pancoast Tumour

Introduction

The thoracic cage, is a bony and cartilaginous structure which surrounds the thoracic cavity and supports the shoulder girdle, forming a core portion of the human skeleton. It also provide support and protection for the heart and lungs.



The lungs are pyramid-shaped, paired organs that are connected to the trachea by the right and left bronchi; on the inferior surface, the lungs are bordered by the diaphragm. The diaphragm is the flat, dome-shaped muscle located at the base of the lungs and thoracic cavity. The lungs are enclosed by the pleurae, which are attached to the mediastinum. The right lung is shorter and wider than the left lung, and the left lung occupies a smaller volume than the right. The **cardiac notch** is an indentation on the surface of the left lung, and it allows space for the heart. The apex of the lung is the superior region, whereas the base is the opposite region near the diaphragm. The costal surface of the lung borders the ribs. The mediastinal surface faces the midline.

The main functions of the lungs are to transfer oxygen from the air to the blood and to release carbon dioxide from the blood to the air.
Air enters the mouth or nose and travels through the windpipe, bronchi and bronchioles to the alveoli. The exchange of oxygen and carbon dioxide takes place in the alveoli:

- The alveoli absorb oxygen from the air and pass it into the blood, which circulates the oxygen around the body.
- Carbon dioxide, which is a waste product of the body's cells, passes from the blood into the alveoli and is breathed out.

The lungs also play a role in the body's defences against harmful substances in the air, such as smoke, pollution, bacteria or viruses. These substances can pass through the nose and become trapped in the lungs. The lungs produce a thick, slippery fluid (mucus), which can trap and partly destroy these substances from the air. The cilia move rapidly to push the mucus up through the bronchi, where it is removed by coughing or swallowing.

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Pancoast Tumour

Pancoast tumours are lung cancers that form at the extreme apex (very top) of either the right or left lung in the superior sulcus (a shallow furrow on the surface of the lung).

Pancoast tells one where the cancer is, rather than what type it is. Pancoast tumours grow right at the top of the lung (the apex). This position makes them rare because most lung cancers develop lower down in the lungs. Fewer than 5 in every 100 cases of lung cancer (5%) are Pancoast tumours.

Because of their location in the apex of the lung, they invade adjoining tissue. They form an abnormal patch of tissue over the lung apex and principally involve the chest wall structures rather than the underlying lung tissue. They invade the following structures:

- Lymphatics (small, thin vessels that carry lymph fluid through the body)
- Lower roots of the brachial plexus (a complex network of nerves that is formed chiefly by the lower 4 cervical [neck] nerves and the first thoracic [chest] nerve)
- Intercostal nerves (nerves that lie between a pair of adjacent ribs)
- Stellate ganglion (a mass of nerve tissue containing nerve cells that form an enlargement on a nerve or on 2 or more nerves at their point of junction or separation)
- Sympathetic chain (either of the pair of ganglionated lengthwise cords of the sympathetic nervous system that are situated on each side of the spinal column)
- Adjacent ribs
- Vertebrae

Most Pancoast tumours are non-small-cell cancers and most commonly squamous cell cancers. Between 35 and 40 out of every 100 lung cancers diagnosed are squamous cell cancers. These cancers develop from the cells that line the airways. Pancoast tumours can be difficult to diagnose.

Gundepalli, S.G. & Tadi, P. 2021 .

“Pancoast or superior sulcus tumor consists of a wide range of tumors invading the apical chest wall and producing a characteristic syndrome named “Pancoast -Tobias syndrome.” The superior sulcus is an anatomical pleuro-pulmonary groove that is formed by the subclavian artery when it curves in front of the pleura and runs upward and lateral immediately below the apex. Not all superior sulcus tumors are in this exact location, but this term signifies any tumor that presents in the apices of the lungs associated with clinical signs and symptoms that are the hallmark of Pancoast syndrome.

The combination of severe shoulder/arm pain along with the distribution of the C8, T1, T2 nerve trunks, Horner syndrome (ipsilateral ptosis, miosis, and anhidrosis; caused due to interruption of sympathetic nerve chain running to the head) and atrophy of the intrinsic hand muscles is termed “Pancoast-Tobias syndrome.”

The overall prognosis of patients with this disease is poor, although recent advancements in the treatment regimen showed significant improvement. Treatment of Pancoast tumors involves interprofessional care coordinated among a thoracic surgeon, a radiation oncologist, and a medical oncologist.”

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Incidence of Pancoast Tumour in South Africa

The outdated South African National Cancer Registry (2017), known for under reporting. does not provide any information regarding the incidence of Pancoast Tumour. According to the outdated National Cancer Registry (2017), known for under reporting, the following number of lung cancer cases was histologically diagnosed in South Africa during 2017:

Group - Males 2017	Actual No of Cases	Estimated Lifetime Risk	Percentage of All Cancers
All males	1 880	1:82	4,70%
Asian males	90	1:65	9,27%
Black males	697	1:147	5,28%
Coloured males	372	1:45	7,89%
White males	721	1:48	3,40%

Group - Females 2017	Actual No of Cases	Estimated Lifetime Risk	Percentage of All Cancers
All females	1 056	1:191	2,53%
Asian females	32	1:266	2,41%
Black females	260	1:560	1,35%
Coloured females	219	1:94	4,81%
White females	545	1:69	3,18%

The frequency of histologically diagnosed cases of lung cancer in South Africa for 2017 was as follows (National Cancer Registry, 2017):

Group - Males 2017	0 – 19 Years	20 – 29 Years	30 – 39 Years	40 – 49 Years	50 – 59 Years	60 – 69 Years	70 – 79 Years	80+ Years
All males	1	5	34	174	518	615	410	123
Asian males	0	1	1	4	14	37	25	8
Black males	1	2	22	91	240	231	90	20
Coloured males	0	0	7	49	115	114	67	20
White males	0	2	4	30	149	233	228	75

Group - Females 2017	0 – 19 Years	20 – 29 Years	30 – 39 Years	40 – 49 Years	50 – 59 Years	60 – 69 Years	70 – 79 Years	80+ Years
All females	1	4	25	65	249	349	268	95
Asian females	0	0	0	5	5	10	7	5
Black females	0	4	13	28	85	73	42	15
Coloured females	1	0	4	14	57	82	48	13
White females	0	0	8	18	102	184	171	62

N.B. In the event that the totals in any of the above tables do not tally, this may be the result of uncertainties as to the age, race or sex of the individual. The totals for 'all males' and 'all females', however, always reflect the correct totals.

Signs and Symptoms of Pancoast Tumour
Because the cancer is at the top of the lungs, it may put pressure on or damage a group of nerves (the brachial plexus) that runs from the upper chest into one's neck, face and arms. This can cause several very specific symptoms:

- Severe pain in the shoulder or the shoulder blade (scapula)
- Pain in the arm and weakness of the hand on the affected side
- Horner's syndrome.

[Picture Credit: Horner's Syndrome]

Horner's syndrome is the medical name for a group of symptoms. One gets flushing on one side of the face and that side does not sweat. The eye on the same side has a smaller (constricted) pupil with a drooping or weak eyelid.

Risk Factors for Pancoast Tumour
The risk factors for almost all lung cancers are similar.

These include:
- Use of tobacco products
- Secondary smoke exposure
- Asbestos exposure
- Exposure to industrial metals such as gold and nickel

Diagnosis of Pancoast Tumour
Imaging and biopsy are the cornerstones of evaluation of Pancoast Tumour. The apex of the lung can be difficult to investigate because it is bounded laterally by the first rib, posteriorly by the first rib and the vertebral bodies, and anteriorly by the costal cartilage of the first rib and the manubrium. Plain radiographs of the chest frequently show no change or an asymmetry or thickening of the apical cap. Apical lordotic films may be more revealing. Computed tomography (CT) and magnetic resonance imaging (MRI) have become standard.

In very rare cases, sputum cytology has been helpful. Initially, most Pancoast tumours are diagnosed histologically on the basis of transthoracic needle biopsy results. Diagnosis via bronchoscopy is less helpful because most of these tumours are peripherally located. The flexible scope is more useful than the rigid scope in obtaining bronchoscopic aspirates and brush biopsy specimens.

Liver, bone, and brain scans are performed to look for metastatic disease. Although more than 90% of patients can be correctly diagnosed on the basis of clinical and radiologic findings alone, open biopsy for pathologic validation may be performed through a supraclavicular incision. Results from a needle biopsy through the supraclavicular or posterior triangle are also successful in confirming the

diagnosis and in delineating the cell type before treatment. Even though clinical diagnosis is relatively simple, performing a tissue biopsy is still necessary.

Issi, Z. & Erkin, Y. 2020.
"A Pancoast or superior sulcus tumor is a rare, bronchogenic carcinoma. In the early period, shoulder pain is the most common symptom. In this case, the patient had presented with complaints of shoulder and arm pain at other outpatient clinics and was examined primarily for musculoskeletal causes and radiculopathy. The patient had no complaints of facial symptoms and Horner's syndrome signs, such as anhidrosis of the face and neck region, were not noticed. Advanced imaging of a patient with preganglionic Horner's syndrome is important. Fewer than 50% of patients with a Pancoast tumor have a resectable lesion at the first diagnosis. Diagnosis is often delayed or there may be a misdiagnosis because musculoskeletal disorders are the focus and there are few lung-related complaints. A detailed examination and anamnesis is very important in patients with arm and shoulder pain."

Laboratory Studies
The blood workup for patients with Pancoast tumours is not specific, and results are not diagnostic.

Lung cancers produce various substances. Elevated levels of oncofoetal carcinoembryonic antigen and beta-2 microglobulins are associated with many lung cancers. Unfortunately, these findings are not diagnostic, because levels of these chemicals are also elevated by other nonspecific causes, such as smoking and bronchitis.

Tumour markers (e.g., bombesin, neuron-specific enolase, and other peptides) are common with small cell cancers and are related to the stage of the disease. They may aid in distinguishing differentiated forms of lung cancer from undifferentiated forms.

Various tumour oncogenes, including K-*ras, c-myc, TP53,* and *HER-2/neu,* have also been identified in patients with lung cancers. Although the presence of these oncogenes has some prognostic value, they are not important for staging of the cancer.

Routine blood work in all patients with a lung cancer usually includes a complete blood count (CBC) count, blood urea nitrogen (BUN) and creatinine levels, a white blood cell (WBC) count, and urinalysis. Coagulation parameters, such as prothrombin time (PT), activated partial thromboplastin time (aPTT), and platelet count, are appropriate. Unless metastatic disease is evident, liver function tests are not regularly performed. Any patient deemed a surgical candidate has blood drawn for a cross-match.

Urinalysis is performed in all patients before surgery, and a catheter specimen is obtained in women if the initial urinalysis result suggests contamination.

Imaging Tests
CT and MRI of the neck, chest, and upper abdomen have largely replaced older radiographic studies in the workup of Pancoast Tumour.

MRI is useful for evaluating resectability. It may be more accurate in evaluating chest wall invasion, examining vascular structures, and assessing the brachial plexus for invasion. It is more accurate than a CT scan for assessing invasion of cervical structures and vertebral bodies.

Additional staging studies should be considered. Mediastinoscopy should be performed to evaluate mediastinal nodes. The presence of N2 mediastinal lymphadenopathy has a significant adverse effect on survival. CT or MRI of the head to exclude occult metastasis should be performed if treatment with curative intent is planned. CT of the chest can be extended to include the liver and adrenal glands.

Chest radiographs may reveal a small homogenous apical cap or pleural thickening; they may show a thin plaque at the lung apex in the area of the superior sulcus or may reveal a large mass, depending on the stage of the tumour when it is first diagnosed. Suggestive films should prompt the astute diagnostician to order apical lordotic views for better visualisation of the area.

Brochoscopy and Biopsy

Bronchoscopy helps evaluate the tracheal and bronchial lumens; however, because most Pancoast tumours are peripheral, the diagnostic yield is low. Whereas sputum cytology results are positive in fewer than 15% of patients, fibreoptic bronchoscopy findings are more often positive - but only in 20-30% of patients, because of the peripheral location of the tumour. Bronchoscopy, however, can be useful in excluding otherwise unsuspected concurrent endobronchial lesions.

Tissue diagnosis is obtained on the basis of results from percutaneous needle biopsy, either under fluoroscopy or under CT guidance. Staging is based on scalene node biopsy results from palpable nodes or mediastinoscopy findings. If a patient presents with supraclavicular lymph node enlargement, then a fine-needle aspiration (FNA) biopsy of enlarged supraclavicular lymph nodes or an ipsilateral supraclavicular fullness procedure is a fast, safe, and inexpensive means of confirming the diagnosis.

Transthoracic needle biopsy by CT guidance has a high yield, up to 95% in some series. Some tumours may be evaluated only by thoracotomy, either open or video assisted.

Other Tests

Rarely, arterial or venous involvement of the subclavian artery or vein occurs; thus, arteriography or phlebography may be helpful. This is usually accomplished in a retrograde fashion, although it can be approached from the opposite extremity or from the leg.

Baseline electrocardiography (ECG) is performed on all patients for comparison to postoperative ECG tracings (if one is performed).

Staging of Pancoast Tumour

The stage of a cancer tells one how big it is and how far it has spread. It is important because it helps the doctor decide which treatment to administer. The tests and scans give some information about the stage. Sometimes it is not possible to be certain about the stage of a cancer until after surgery.

Treatment of Pancoast Tumour

Originally, Pancoast tumour was fatal due to involvement of vital structures at the thoracic inlet. This has improved with multimodality treatment, including induction chemoradiotherapy (usually cisplatin-based) followed by resection. Resection may involve a wedge resection or a lobectomy (surgical removal of a whole lobe of the lung). Traditionally, the involved brachial plexus was also resected, leading to paralysis and neuropathic pain.

Involvement of vertebrae, cervical plexus and lymph nodes are all associated with poorer outcomes. Historically, five-year survival was 30-40% with complete resection and no lymph node involvement and <10% for all other groups. Two-thirds of patients experience a recurrence. However, data regarding survival rates with induction chemoradiation and resection are much better and five-year survival rates of approximately 50-70% have been reported. Mediastinal lymph node involvement is associated with a particularly poor prognosis.

Shimada, Y., Kudo, Y., Maehara, S., Hagiwara, M., Tanaka, T., Shigefuku, S., Kakihana, M., Kajiwara, N., Ohira, T. & Iked,a N. 2020.

Objective: In this study, we aimed to identify prognostic determinants and to comparably analyze clinical features of patients with both resected and unresected superior sulcus tumors (SSTs).

Methods: The data of 56 patients who underwent any treatment for an SST from 2004 through 2016 in our hospital were reviewed. Overall survival (OS) rates were estimated using the Kaplan-Meier method. Univariate and multivariate analyses were performed to determine independent prognostic factors for patients with resected and unresected SST separately.

Results: The number of patients with resected and unresected SSTs was 24 (43%) and 32 (57%), respectively. Of the 24 patients who underwent surgery, 20 received induction therapy, with 32% achieving pathological complete response. Complete resection (R0) was performed in 22 patients (92%). On multivariate survival analysis, preoperative serum carcinoembryonic antigen (CEA) level (median 8.3 ng/ml, p = 0.021) was identified as the independent determinant of OS in surgical patients; whereas, initial treatment response (complete response or partial response, p = 0.032) was the independent OS indicator in non-surgical patients. The 5-year OS of the patient with resected and unresected SST was 68.8% and 29.1% (p = 0.008), respectively.

Conclusion: Significant prognostic factors differ among patients stratified by the presence of surgical resection for SSTs. Preoperative CEA level in surgical candidates and initial treatment response in non-surgical patients were the independent factors associated with OS. Surgical candidates are expected to have more favorable survival than patients with unresectable SSTs.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details. For additional information, please visit: www.sanctr.gov.za/

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