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Pancoast's syndrome

Synonyms: Tobias syndrome, Ciuffini-Pancoast-Tobias syndrome

What is Pancoast's syndrome?[1]

Pancoast's syndrome is usually caused by a malignant neoplasm of the superior sulcus of the lung which produces destructive lesions of thoracic inlet along with involvement of brachial plexus and stellate ganglion. These cancers are rare, accounting for fewer than 5% of all lung cancers. The neoplasm is usually bronchogenic in origin (about half are squamous cell carcinomas, half adenocarcinomas). [2]

Pancoast's syndrome symptoms (presentation)^[2]

This syndrome results from the invasion of a number of structures and tissues around the thoracic inlet and may be characterised by:

- Ipsilateral invasion of the cervical sympathetic plexus, leading to Horner's syndrome (miosis, enophthalmos, ptosis) in 14-50% of patients. [3]
- Ipsilateral reflex sympathetic dystrophy in the arm, with enhanced sensitivity to touch, and skin changes.
- Shoulder and arm pain (from the brachial plexus invasion C8-T1) with wasting of the intrinsic hand muscles and paraesthesiae in the medial side of the arm.
- Less commonly, unilateral recurrent laryngeal nerve palsy producing unilateral vocal cord paralysis (hoarse voice ± bovine cough), and/or phrenic nerve involvement.
- Arm oedema secondary to the compression of blood vessels.
- Superior vena cava obstruction, with facial swelling, difficulty breathing and a flushed appearance.

The main presenting features of shoulder and arm pain, in the absence of respiratory symptoms, can result in a delay in diagnosis while the patient is inappropriately investigated for cervical arthritis, shoulder bursitis and rotator cuff injuries. [4]



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Other causes

Malignancies other than lung cancer can result in Pancoast's syndrome. Primary cancers have been reported, such as breast, mesothelioma, plasmacytoma or lymphoma; ^[5] or metastatic carcinoma from the larynx, cervix, bladder, thyroid or colon. ^[6]

Non-neoplastic causes of Pancoast's syndrome are rare. However, there have been reported cases due to bacterial pneumonia (staphylococcal or pseudomonas), tuberculosis, hydatid disease, [7] [8] mycotic aneurysm, disseminated nocardiosis (a rare Gram-positive bacterial infection), plasma-cell granulomas, a cervical rib and pulmonary amyloidosis.

Staging

This is as for other bronchogenic carcinomas, ie the tumour, node and metastasis (TNM) classification. Most Pancoast tumours are T3 at presentation, as there is invasion of pleura and the brachial plexus. [3]

Investigations^[1]

This is similar to other lung cancers with an emphasis on imaging, including CXR and CT scan of the lungs and abdomen, and also possibly positron emission tomography (PET). MRI is the imaging of choice to assess structures at the thoracic inlet prior to surgery. [3] [9] Brain CT or MRI are usually performed, as it is the most common site of metastases. Good biopsy results are usually achieved by percutaneous methods. [10]

Pancoast's syndrome treatment and management [1] [11]

The National Institute for Health and Care Excellence (NICE) recommends that Pancoast tumours should be treated in the same way as other types of non-small cell lung cancer (NSCLC), with multimodality therapy according to resectability, stage of the tumour and performance status of the person. [12]

However, management may differ from other types of non-small cell lung cancer (NSCLC). Its position and close proximity to vital structures may make surgery difficult. Therefore, depending on the stage of the cancer, treatment may involve radiation and chemotherapy given prior to surgery.

Surgery may consist of the removal of the upper lobe of a lung together with its associated structures as well as mediastinal lymphadenectomy.

There are a number of surgical approaches, each with their advantages and disadvantages. ^[13] The three most common procedures are the high postero-lateral approach, the anterior transcervical thoracic approach and the anterior trans-sternal approach.

Resection may involve a wedge resection or a lobectomy. [3] [10] [14] Historically the involved brachial plexus has also been resected, leading to paralysis and neuropathic pain; however, this has largely been shown to be unnecessary. [15]

Prognosis

Until the 1990s bimodal treatment (pre-operative radiotherapy followed by surgery) resulted in a five-year survival at best of only 30%, and a recurrence rate of 70%. [13] However, the advent of trimodal therapy (induction chemoradiotherapy followed by resection) has decreased recurrence rates to 30%; five-year survival remains modest at between 40% and 70%. [3] [13]

Further reading

- Villgran VD, Chakraborty RK, Cherian SV; Pancoast Syndrome. StatPearls, Jan 2023.
- Gundepalli SG, Tadi P; Lung Pancoast Tumor. StatPearls, Jan 2023.

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