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CASE REPORT OPEN ACCESS

PANCOAST TUMOR PRESENTING WITH HORNER SYNDROME: A CASE REPORT

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ABSTRACT

Case Report: A 50-year-old man was referred to Dr. Moewardi General Hospital with history of right chest pain that radiate to the right back and hand for 2 months. His complaint did not improve with medication and physical therapy. On physical examination we observed multiple nodules on the right axilla, scapula, and colli region. Neurological examination revealed right sided partial ptosis, anisocoria, ipsilateral weakness, and ipsilateral allodynia, and anhidrosis T1 dermatome. CT-scan showed a right lung mass in the superior lobe of the apical segment. The diagnosis of Horner syndrome is then established caused by multiple lymphadenopathies. His symptoms then relieved after lymphadenectomy.
Conclusion: Horner syndrome, characterized by ipsilateral pupillary anisocoria, ptosis and anhidrosis, is one of the complications of Pancoast tumor invasion to the sympathetic nervous system in the C8, T1, and T2 nerve territory.

Keywords: Pancoast tumor, horner syndrome, case report

Introduction

Pancoast tumor, which comprises a variety of tumor on lung superior sulcus, could invade the apical chest wall and produce a characteristic syndrome commonly known as Pancoast syndrome. Characteristics of this syndrome include ipsilateral shoulder and arm pain in the distribution of C8, T1, and T2 nerves, paresthesia, paresis, hand thenar muscle atrophy, and Horner syndrome (ptosis, miosis, and anhidrosis)[1,2]. Pancoast tumors cause second-order or pre-ganglionic Horner syndrome.[3]

Pancoast tumors usually cause neurological disorders involving the brachial plexus causing shoulder and arm pain as well as Horner syndrome. Musculoskeletal symptoms such as chest pain is conceivable if the parietal pleura, ribs, or vertebral bodies are involved.[3]

Pancoast tumor has no specific laboratory examination for it is a group of malignancies classified by location. A chest X-ray will show soft tissue opacity at the lung apex.

A CT scan could provide more information about the extent of the primary tumor, mediastinal lymphadenopathy, satellite tumors, and is magnificent to identify bone involvement. MRI is used to determine staging due to it can depict involvement of lung tissue, blood vessels, spinal cord and brachial plexus invasion.[1]

Case Report

A 50-years-old man was referred to neurology clinic in Dr. Moewardi General Hospital, Surakarta. Patient's major complaint was right chest pain radiating to the back and upper right arm for 2 months. The pain progressively and continuously felt then spread to the fingers in the right hand. Ptosis was found. The pain was described as an electric shock and worsen with light touch. The patient also had painless lumps in the right neck and back.

Clinical Findings

Initial examination showed right anterior chest pain with Visual Analog Scale score of 6. The patient obeyed commands and good oriented. The airway was clear, respiratory rate 20 times per minute, heart rate 78 beats per minute with regular rhythm, and blood pressure was 130/80 mmHg.

Patient's Glasgow coma scale score was 15 (E4V5M6). General physical examination revealed an anisocoric pupil with diameter of 2mm/4mm, normal light reflex, and right partial ptosis (Figure 1). Lumps were found the right axilla (diameter ±1 cm), right scapula (diameter ±2 cm), and right coli region (diameter ±2 cm). From neurological examination, motoric evaluation showed the right upper arm strength was decreased with muscle strength of 444. Muscle tone in all four extremities and physiological reflexes were normal. There were no pathological reflexes found. Allodynia was found in the right upper extremity. Ciliospinal reflex was found negative in the right eye, and positive in the other. We also set a further test to figure out whether the anisocoric pupil was truly evolved by sympathetic disturbance or other causes. Brimonidine drops (α-2 adrenergic agonist) were used to replace apraclonidine, the test revealed an insignificant result.

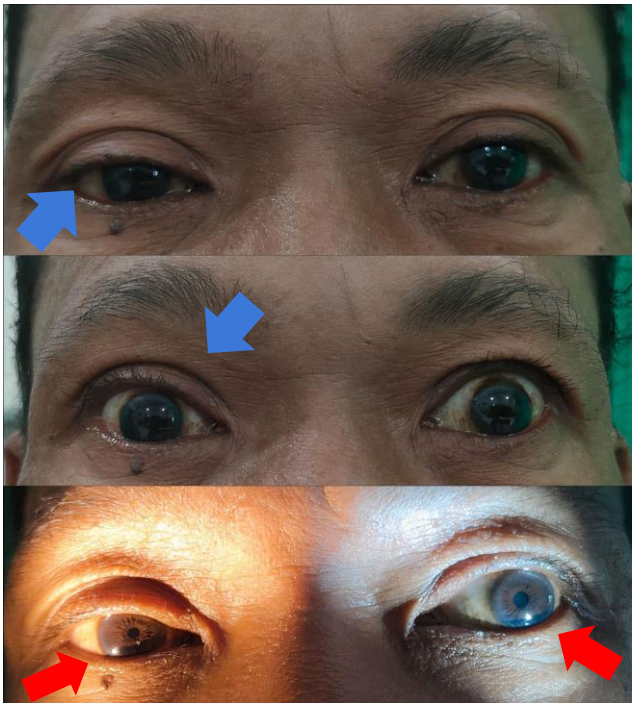

Figure 1. Ophthalmic Examination. **A.** Partial ptosis (blue arrow). **B.** Anisocoric pupil (red arrow)

A perspiration test was performed and showed that the patient's autonomic function was still good throughout the body, except for the T1 dermatome area.

The chest X-ray showed an inhomogeneous opacity on the right superior aspect of the hemithorax, suspected lung mass with pleural mass, and pneumonic type pulmonary metastases (Figure 2).

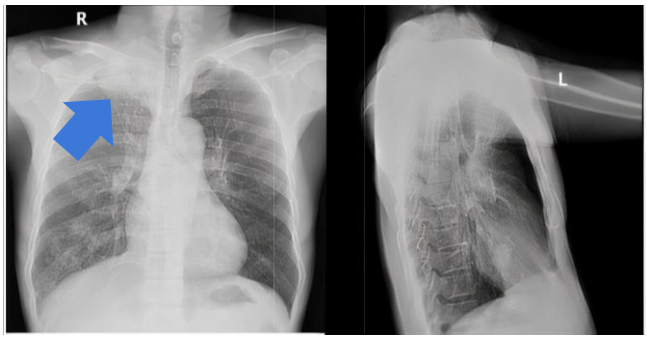

Figure 2. Chest X-Ray, inhomogeneous opacity on the right superior aspect of the hemithorax

Chest MSCT-Scan with contrast showed a right lung mass in the superior lobe of the apical segment with a pneumonic reaction attached to the superior vena cava, right visceral pleura, right superior and anterior chest wall. Multiple lymphadenopathies were found in left mediastinum V-VII, bilateral axillary, supraclavicular and bilateral infraclavicular. The staging according to the AJCC VIII edition in 2017 is T3N3M0, which is stage IIIC (Figure 2).

Painkillers such as paracetamol, tramadol, diazepam, and amitriptyline were given. A right-coli lymphadenectomy was performed on the 5th day. The lymphadenectomy procedure obtained 2 pieces of white-brownish tissue, with 2 cm and 0.5 cm diameter. On microscopic evaluation, lymph node tissue was found with tumor nests composed of solid cribriform, tubular cells consisting of heavy pleomorphic cells, scanty cytoplasm, nucleus with coarse chromatin, and prominent nucleoli with mitosis. This finding indicates adenocarcinoma tissue metastasized from the Pancoast tumor. After lymphadenectomy, he affirmed that the pain, allodynia, and ptosis were diminished.

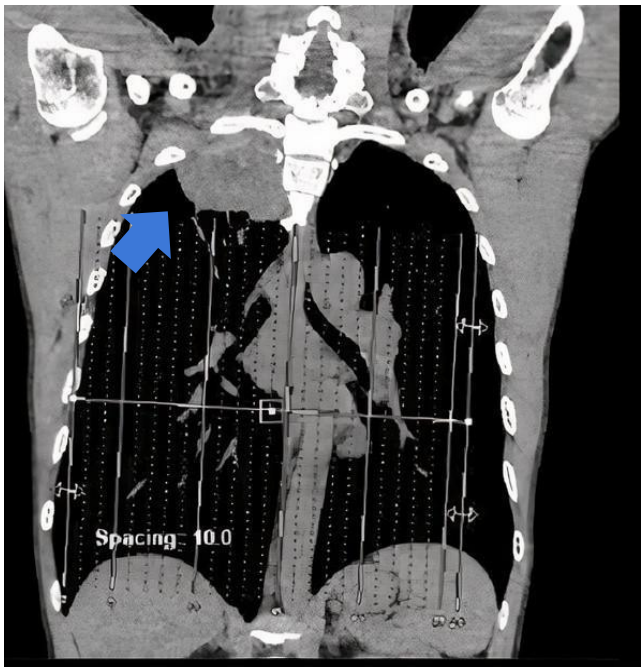

Figure 3. Coronal contrast CT-Scan, solid lesion with spiculated irregular edges in the right apical segment of the superior lobe of the lung

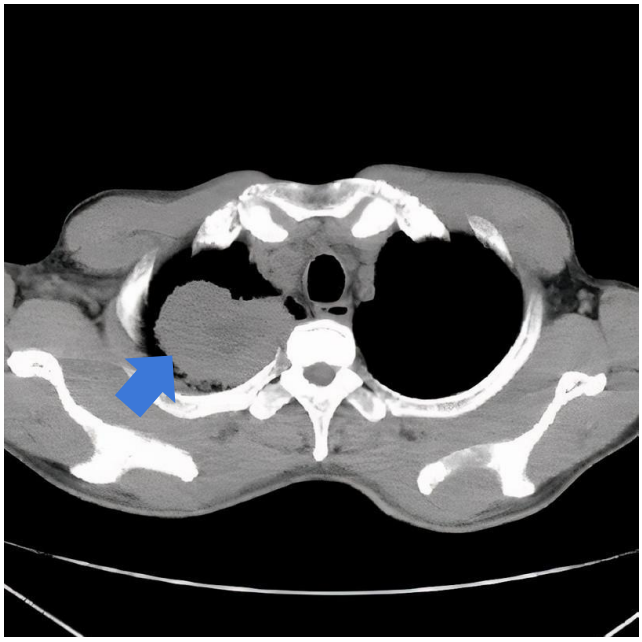

Figure 4. Axial contrast CT-Scan, longest axis of the lesion is 5 cm

Discussion

Pancoast tumor is a lung tumor located at the apex of the lung and is one of the rare cases of tumor.[3] The prevalence of this type of tumor is only 3-5% of all lung carcinomas.[3,1] Invasion and enlargement of Pancoast tumor will compress surrounding organs such as the brachial plexus, sympathetic nervous system, and or spinal nerves which causes signs and symptoms based on the location of the invasion. [1]

Lung cancer has various clinical manifestations, which can be intrathoracic tumor effects, extra thoracic spread, or

paraneoplastic phenomena.[5] To diagnose Pancoast tumor is quite difficult since the initial symptoms resemble musculoskeletal symptoms which only pain or weakness in the upper extremities,[1] make the symptoms often misinterpreted as musculoskeletal problem.

In this case, the initial symptoms were chest pain and right shoulder pain radiating to the distal right hand. The pain worsened for the last 2 months, along with the appearing of some lumps/mass in his right neck and axilla region. These complaints are due to tumor invasion reaching the brachial plexus, which regulates motor and sensory innervation of the upper extremities, along with shoulders, arms, and hands[3,1]

History of smoking and male gender were found as risk factors in this patient. The risk factors associated with Pancoast tumor are the same as lung cancer. Nowadays, smoking is the most common risk factor. Besides, passive smoke inhalation, work-related exposure of industrial element, infection such as TB, age in sixth decade, and genetic are also important.[3,5]

MSCT radiology results showed a right lung mass attached to the superior vena cava, right visceral pleura, right superior and anterior chest wall. Despite this finding, the increasing pressure of superior vena cava symptoms such as flushed appearance, edema of the face, and right superior extremity were not found in this patient.

Allodynia

Allodynia, a neuropathic pain caused by a stimulus that should not induce pain perception was also felt in this patient.[6]

Allodynia occurs due to excessive mechanosensitive of nociceptive sensory nerve endings that are processed by spinal cord circuits. On Pancoast tumor, a spike in the activity of A-beta fibers was found and sensitizing the central and/or peripheral nervous systems. The presence of spontaneous ectopia in the periphery will induce and maintain central sensitization.[7] In this case, brachial plexus sensitization was caused by suppression of the Pancoast tumor as spontaneous ectopia.

Ciliospinal Reflex

Ciliospinal reflex is a pupillary dilation reflex secondary to sympathetic activation, caused by noxious cutaneous stimulation of ipsilateral head, neck or upper trunk.[8] The patient showed negative ciliospinal reflex in the right eye, it confirmed the sympathetic disturbance ipsilateral to the side of Pancoast tumor.

Horner Syndrome

Horner syndrome is a rare condition characterized by partial ptosis (dropping of the upper eyelid), miosis (constricted pupils), and facial anhidrosis (absence of sweating) due to disruption of the sympathetic nerve supply.[9]

Perspiration test is done to evaluate the function of the autonomic nerves, as well as to determine the height and location of the lesion in the spinal cord.[4] Perspiration process is regulated by sympathetic nervous system.[10] The test performed in patient showed sweat production disturbance along T1 dermatome area. This finding suggests the T1 sympathetic nervous system involvement.

The ophthalmic examination to this patient showed anisocoria pupil dilation and right partial ptosis. These symptoms arise due to sympathetic pathway disruption.[9] Partial ptosis in Horner syndrome occurs due to disturbances in the oculo-sympathetic pathway causing paralysis of the dilator pupillae and Müller muscles which causes miosis and ptosis.[9,11] The sympathetic nervous system is responsible for pupillary dilation, pupillary constriction in Horner syndrome is due to the dominant parasympathetic nervous system. Pupillary light reflex and accommodation are normal because these reflexes do not involved in sympathetic pathways.[9]

To confirm Horner syndrome, an α-agonist agent is needed. Patients with Horner syndrome have denervation hypersensitivity of the α-1 receptors in the iris stroma of the affected eye.[12] Alpha-agonist can be applied to both eyes to observe the reversal of miosis on the affected side.[13] Alpha-agonist agent used to confirm Horner syndrome is Apraclonidine, but it was unavailable in our country, so we tried another α-agonis agent, brimonidine.

However, the result was insignificant. Brimonidine is a third-generation alpha-agonist which has both α-2 and α-1 adrenoceptor effect but dominantly act as a highly α-2 selective adrenoceptor agonist.[12,14] Meanwhile, apraclonidine has also α-2 and a weak α-1 receptors reaction, which gives minimal to no clinical effect on the pupils of normal eyes and gives significant reaction to the affected eyes in Horner syndrome.[12] This argument suggests that brimonidine should not be used as the confirmation test for Horner syndrome.

Furthermore, anhidrosis on the ipsilateral side is also found in this patient.[9,14] The presentation of anhidrosis depends on the degree of sympathetic nervous system disturbance. First-order lesions affect the ipsilateral side of the body, second-order lesions affect the ipsilateral face, and third-order lesions are limited to the medial side of the face[10,16].

Chemoradiation and surgical resection are the mainstay of therapy for Pancoast tumors. Contraindications to Pancoast tumor resection surgery include: metastases, mediastinal or supraclavicular gland involvement, >50% vertebral body involvement, esophagus and/or trachea, and brachial plexus above T1.[1] Pancoast's tumor in this case was not operated because the patient already had multiple lymphadenopathy in mediastinum and supraclavicular gland with clinical symptoms which suggested the involvement of brachial plexus.

A study revealed, the average survival time for Pancoast tumor received chemoradiotherapy is ±2-5years.[16] The lymph node involvement may presume the prognosis, N2 staging particularly associated with 2-years relapse.[16]

Conclusion

Horner syndrome is one of the complications of Pancoast tumor invasion to the sympathetic nervous system along C8, T1, and T2 levels. This syndrome is characterized by ipsilateral pupillary anisocoria, ptosis and anhidrosis to the site of tumor invasion.

Authors suggest that Pancoast tumor needs to be considered in middle-aged men, with history of smoking and complaining of chest and shoulder pain, neurological

symptoms in upper extremity, and Horner syndrome, but have no risk factors at work or a history of previous trauma. This study also revealed that Brimonidine should not be used to confirm Horner syndrome.

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Conflict of Interest

None.

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