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Authors

Izuchukwu, Ifeoma S. Towfigh, Ali

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CLINICAL VIGNETTE

A Case of Undifferentiated Carcinoma Presenting with Extensive Neurological Deficit and Cord Compression

Ifeoma S. Izuchukwu, M.D. and Ali Towfigh, M.D.

A 58-year-old black male presents with right shoulder pain of 5 months duration. The patient describes pain as 10/10, starting in severity at the shoulder blade and radiating to the fingers. There was associated numbness, tingling and heaviness of the whole right arm. He had seen three previous physicians for same pain and had been prescribed physical therapy and medications that were not effective. Medications included methocarbamol, tramadol, menthol, and gabapentin. Past medical history includes polysubstance abuse, tobacco use disorder, reflex sympathetic dystrophy, hepatitis C and prostate cancer diagnosed two years ago. He was homeless and did not have a consistent primary care provider.

On physical examination, the patient was in moderate to severe pain, holding his right hand. Vital signs were normal except for a pulse of 112/min. He could not make a fist with his right hand; the right hand was obviously more swollen than the left hand. It was difficult to examine him due to the amount of pain, numbness and tingling of the right upper extremity.

Right ptosis was noted but pupils were equal, round and sluggishly reactive. Visual fields were full to confrontation, there was left internuclear ophthalmoplegia (when looking to the right, patient's left eye did not adduct completely), brisk right beating nystagmus in both eyes, more brisk when looking to the right; otherwise, extra ocular muscles were intact. His face was symmetric with good forehead wrinkle and smile excursion bilaterally. Palate elevates equally bilaterally. Normal strength Supraclavicular muscle bilaterally and left trapezius. Right Upper Extremity (RUE) movement limited by pain but right trapezius had reduced strength. Tongue was midline.

MOTOR: Exam revealed diminished bulk throughout. Muscle tone difficult to assess in RUE due to pain but tone seemed normal Bilateral Upper Extremity (BLE); however there was flaccid tone in the Bilateral Lower Extremity. He had trace movements of the fingers but would not move his RUE otherwise due to pain.

SENSORY: Exam was remarkable for a T_3 - T_4 sensory level to pinprick testing and cold sensation. No distinct sensory level detected on vibratory sensation testing but there is a gradient with improving vibratory sensation farther up the body.

Deep Tendon Reflexes: The right and left, triceps, biceps and brachial-radialis reflexes were present, absent ankles and toes downwards. Negative Hoffman bilaterally. Negative abdominal reflex

Laboratory work up was remarkable for a chest radiography showing significant increase in parenchymal opacity in the right apex.

Computerized tomography of the chest showed large soft tissue mass in the apical segment of the right upper lobe partially destroying right first and second ribs invading the right upper chest wall. This is associated with significant right supraclavicular, possible right axillary adenopathy and tumor invasion of the right axillary, right subclavian and right brachiocephalic vein. Tumor imaging showed an intensely hypermetabolic mass occupying the upper right lung and extending into the mediastinum downward to the level of the carina, into the chest wall and into the soft tissues of the neck upward to the level of Cervical 7. Cervical 7, Thoracic 1, 2, 3 and 4 vertebra, as well as first and second ribs. (Image 1)

Right posterior neck biopsy showed undifferentiated carcinoma with focal neuroendocrine features, lymph node. Immunohistochemistry showed Pancytokeratin, CK7, CK20, TTF, CD45, CD3/CD20, S100, PSA, Chromogranin are negative. Vimentin was positive. Synaptophysin was focally positive.

The patient developed flaccid paralysis and areflexia of the bilateral lower extremities. He had slightly diminished vibratory sensation in the lower extremities and impaired proprioception of the left great toe. This was secondary to cervical/thoracic cord compression following expansion of the right apical lung mass into the epidural space encasing the spinal cord from C6-T3. He died following refusal of cord decompression and first dose of chemotherapy.

Discussion:

A Pancoast tumor, or superior sulcus tumor, is named after Henry Pancoast, a US radiologist who described them in 1924 and 1932. It is a tumor of the pulmonary apex situated at the top end of either lung in the superior sulcus. The tumor can cause compression of nearby vital structures causing a constellation of symptoms such as Pancoast syndrome (includes Horner's syndrome) and superior vena cava syndrome¹. It is typically found in conjunction with a smoking history and most are non-small cell lung cancers.

Aside from general symptoms such as malaise, fever, weight loss and fatigue, a pancoast tumor can involve the brachial plexus causing pain and weakness in the muscles of the arm and hand. Involvement of the paravertebral sympathetic chain and the inferior cervical (stellate) ganglion leads to Horner's syndrome on the affected side resulting in miosis (constriction of the pupils), anhidrosis (lack of sweating), and ptosis (drooping of the eyelid). The tumor can also compress the right recurrent laryngeal nerve causing hoarseness of the voice. In superior vena cava syndrome, obstruction of the superior vena cava by a tumor's mass effect causes facial swelling, cyanosis and dilatation of the veins of the head and neck.

The clinical signs and symptoms can be confused with neurovascular compromise at the level of the thoracic outlet or cervical radiculopathy, which in the latter case, can lead to a delay in diagnosis. The patient's smoking history, rapid onset of clinical signs and symptoms, and pleuritic pain raised suspicion for an apical tumor.

The treatment of a Pancoast lung cancer can be unique from other types of non-small cell lung cancers owing to its position and close proximity to vital structures (such as nerves and spine) which may make surgery difficult. As a result, and depending on the stage of the cancer, treatment may involve radiation and chemotherapy given prior to surgery (neoadjuvant treatment)². Surgery may consist of the removal of the upper lobe of a lung together with its associated structures (subclavian artery, vein, branches of the brachial plexus, ribs and vertebral bodies), as well as mediastinal lymphadenectomy.

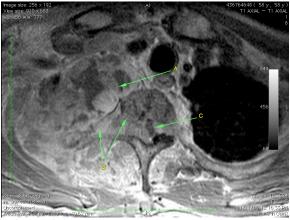


FIGURE 1

Lung mass showing invasion of the vertebra, epidural space and spinal cord.

A is the lung mass

B is invasion of a rib and vertebral body

C is the spinal cord being compressed by tumor

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