CASE REPORT

A case study and literature review of surgical treatment for brachial plexus pain caused by Pancoast syndrome

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Abstract: This study reports the diagnosis and treatment of brachial plexus-associated intractable pain caused by Pancoast syndrome, along with the associated literature review, based on a patient we attended to in April 2011. A full examination was performed after his admission, and neurolysis was performed on the right brachial plexus of lower trunk. Follow-ups and treatment evaluations were carried out 1, 2, 3, and 6 months after the initial neurolysis procedure, which was followed by a radiation therapy two months later. The patient’s pain symptoms were relieved and he partially regained the sense awareness and movement of his right hand. We found that early clinical manifestations of Pancoast syndrome are atypical and are sometimes difficult to detect. It is very similar to brachial plexus injury-related pain, and the patient must be referred to an orthopaedics or hand surgery specialist for treatment. Therefore, improving medical practitioners’ general understanding of this disease is essential in order to avoid potential misdiagnoses.

Keywords: Pancoast syndrome; Horner syndrome; diagnosis; treatment


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Introduction

Pancoast syndrome (or Pancoast’s syndrome) is a clinical syndrome that attacks the lower trunk of brachial plexus with malignant neoplasm[1]. It is also synonymous with superior pulmonary sulcus syndrome, lung cancer, superior pulmonary sulcus tumour syndrome, and superior pulmonary sulcus tumour. It comprises 3%–5% of the total lung cancer incidence rate[2]. In the early stages, the main manifestations of Pancoast syndrome include persistent severe pain in the shoulder, armpit, and arms, or progressively worse spreading pain in the arms, as well as ipsilateral Horner syndrome.

During disease progression, muscular atrophy will affect the arms, thaner, and hypothener, leading to hand dysfunction. Symptoms affecting the lower trunk of brachial plexus (C8-T1 nerve roots) are typically the main reason why patients consult hand surgery specialists or orthopaedists[3]. As the area is distinct and clinical manifestations are non-specific, there is a high misdiagnosis rate[4] of up to 86.9%[5]. Most patients are already in the latter stages when accurately diagnosed, and treatments are usually ineffective. Herein, an analysis of a patient diagnosed with Pancoast syndrome is reported.

Case report

A 40-year-old male patient complained of experiencing pain on his right arm, which has been dysfunctional for more than four months. His condition deteriorated into intractable pain in the hand and subsequently, he was unable to sleep for months. He was then admitted to the hospital on 18th April, 2011 for “unknown origin of pain in the right brachial plexus”.

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Owing to a right middle lobe adenocarcinoma, he previously underwent a resection for right lung adenocarcinoma and a lymphadenectomy for the upper part of his right clavicle in April 2010 at another hospital. After receiving six rounds of adjuvant chemotherapy (particulars unknown), the patient showed a good recovery. Four months later, the patient felt pain and discomfort after performing heavy lifting; however, owing to his generally good movement conditions, his initial complaints were not followed through. Thereafter, the symptoms developed chronically. The pain, especially paralysis in the ulnar skin and a burning sensation, spread through his whole right arm within a month, causing limited hand flexion and was seriously affecting the patient’s sleep quality at night.

The effects of prescribing nutritional intervention targeted at the nervous system and analgesic therapy were insignificant. Moreover, a body check showed an acute haggard face, with the right side being positive for Horner syndrome. A 30-cm long surgical scar was seen along the intercostal space at the centre of his right chest. An approximately 5-cm long surgical scar was seen crossing his right clavicle. The right forearm was thinner than the left forearm, revealing obvious muscular atrophy. Both the thenar and hypothenar of his right hand were evidently atrophying. Besides that, he was also experiencing hypoesthesia in the right forearm ulna, right little finger, ulnar half of the right ring finger, and part of the hand between the thumb and forefinger. Meanwhile, his right wrist flexion strength was at Grade 3, whereas his right finger flexion strength was at Grade 0. Tinel’s test was positive for the upper part of his right clavicle and ulnar nerves of arm, where the parts were out of shape.

Auxiliary examination using electromyography showed a wide motor unit action potential (MUAP) for the right C8-T1 innervation of forearm muscles, decreased amplitude of right median nerve conduction, slowing down of conduction velocity, obvious decrease in the amplitude of right ulnar nerve conduction (along with the slowing down of conduction velocity), thus suggesting that the right C8-T1 had neurogenic damage with electrophysiological characteristics. B-scan ultrasonography of brachial plexus (carried out on 14th April, 2011) showed tumour-like change in the right brachial plexus C8 towards intervertebral foramen, and the T1 nerve was suspiciously thickened, conditions which are considered as lower trunk injury (formation of traumatic neuroma).

After admission, the patient was diagnosed with: (1) traumatic brachial plexus neuroma, and (2) Pancoast syndrome. After the patient and his family were given the details of his condition, a neurolysis procedure was performed on his right brachial plexus. During his surgery, severe muscle fibrosis was found 3 cm next to the site of anterior scalene, which had formed a compact cord-like ligament (Figure 1), oppressing the artery and lower trunk of brachial plexus. After removing all the compact cord-like ligament of anterior scalene, the artery below the clavicle rose by about 0.5 cm. To protect the artery, which is below the clavicle, the lower trunk nerve was carefully separated from the surrounding tissue, the artery below the clavicle, and the lower trunks. Electrophysiology with a super current of 80 mA was administered at the lower trunk of brachial plexus for 5 min. A negative pressure aspirator was placed within after it stopped bleeding. The surgical incision was then sutured layer-by-layer.

![Figure 1. Scarring on the anterior scalene of brachial plexus changed the appearance of the ligament](image)

Post-surgical care and assessment included nutritional therapy that was given after the surgery, administration of antibiotics to prevent infection, as well as the appropriate measurement of fluid infusion. Suture removal was performed 10 days after the surgery, and the patient proceeded to receive moderate functional training. Subsequent evaluations showed that the patient’s pain was relieved on the day after surgery; the pain in the forearm was gone and the patient claimed that he could finally sleep peacefully. His arm’s sense awareness and movement had slightly improved. However, the pain in his arm (at the same area) recurred two weeks after surgery, though not as severe as previously described and paracetamol and tramadol hydrochloride tablets were effective for killing the pain.

Two months after surgery, the patient claimed that his pain had improved, and he was able to sleep better. Nonetheless, based on the magnetic resonance imaging (MRI) of his right lung that was performed subsequently, a high brightness shadow was observed in the apex of his lung. Thus, the patient was subjected to radiation therapy in his right clavicle, which further eased his pain. The
pain in the patient’s right arm was gone three months after his surgery but he complained of a burning sensation akin to his skin being placed on a coal cinder. Unfortunately, the sense awareness and movement of his upper right arm showed no obvious improvement. The patient experienced swelling and discomfort in his arm, especially at the back of his hand, six months after his surgery. Since the onset of the disease, Horner syndrome continued to develop on the patient’s face. According to the evaluation standards for the lower trunk of brachial plexus, the patient’s conditions (i.e. before and after surgery) were generally poor. A follow-up three years after the patient’s initial surgery confirmed that the patient’s sense awareness and movement in his arm had no obvious improvements, and he eventually died due to metastatic carcinoma.

Discussion

The earliest report of disease characteristics alluding to Pancoast syndrome was described in 1838 by an English surgeon, Edward Selleck Hare. In 1924, American radiologist Henry K. Pancoast published further reports of this disease, describing three major manifestations that are associated with the disease: severe pain from shoulder to hand, muscular atrophy in the wrist and hand, and ipsilateral Horner syndrome. Eight years later, Pancoast came out with an additional report of seven patients with Pancoast syndrome, and officially named this disease as superior pulmonary sulcus tumour. The pathology was divided into two categories: (1) tumours in the apex of lung (including primary tumour in apex of lung such as primary lung cancer, cervical spine tumour, laryngeal cancer, Hodgkin’s disease, and pleural mesothelioma, among other things), and metastases from sites of origin such as stomach, pancreas, thyroid, breast, oesophagus, etc.; (2) other non-malignant lesions such as benign tumour and injury, as well as non-specific and specific inflammations, such as tuberculosis, hydatid cysts, etc. The trait of this disease that causes inflammation is being taken seriously by experts; however, the pathology of primary tumour in apical site still remains dominant.

Clinical manifestation: In our patient, apical lesion was evident but he lacked certain lung cancer characteristics such as cough, difficulty to breathe, and hemoptysis, among other things. His conditions were deemed similar to neuropathic pain caused by brachial plexus injury and thus, this became the main reason of misdiagnosis. Other possible clinical manifestations include: (1) symptoms such as apical or pleural malignant cancer, neck and shoulder pain, and pain radiating up to the head, down to the scapular, axilla, chest, ulnar nerve area, etc.; (2) pain in the shoulder and axilla as described above, with a persistent severe pain in the upper limbs that worsens and spreads, and slowly leading to paralysis in the ipsilateral arms, especially in the thenar and hypothenar muscles; (3) showing ipsilateral Horner syndrome; and (4) general weakness and cachexia in patients.

Diagnosis: (1) Clinical manifestation. When patients have symptoms such as neck and shoulder pain, along with a pain that radiates to the forearm, the onset of this disease should be considered, especially for patients with a history of lung cancer. As the disease progresses, the appearance of Horner syndrome, severe pain or paralysis in the forearm, and a simultaneous muscular atrophy in the hand should alert the doctors of this disease; however, it should be differentiated from a hand surgery’s thoracic outlet syndrome, dorsal scapular nerve compression, and brachial plexus avulsion pain. (2) Imaging examination. (a) Lateral chest X-ray is the preferred method and the reference point of other imaging procedures. As high as 90% of apical lung carcinoma patients show abnormal shadow in the apex of lung. In the early days, chest X-rays were less effective owing to a multitude of reasons, which contributed to unnecessary delays in diagnosis and treatment. Nonetheless, improvements in technology and skills have greatly increased their accuracy. (b) Chest computed tomography (CT) scan is useful for the diagnosis of superior pulmonary sulcus tumours in their early stages, as it is able to show a clearer image of any thickening parts, nodules, and lumps of apical pleura. This is helpful in differentiating superior pulmonary sulcus tumour from other apex solitary nodules or lumps. (c) Magnetic resonance imaging (MRI) scan is helpful in providing a significantly clearer relationship between the tumour, blood vessels, and nerves in the process of establishing the extent of tumour invasion to the rib, head, and spinal vertebrae, as well as to determine if the tumour had violated the spinal vertebrae, i.e., evaluating the level of invasion to the brachial plexus.

Using imaging examination, it was discovered that by studying apical lesions and the adhesion, infiltration, and destruction of the surrounding tissue, doctors are able to diagnose this disease more accurately. In addition, MRI also helps to guide the surgical procedure as it evaluates the level of invasion of superior pulmonary sulcus tumour into all thoracic inlet structures, which include the brachial plexus, the artery below clavicle, vertebra, nerve roots, and foramen. (d) B-scan ultrasonography for brachial plexus is a convenient and intuitive low-cost imaging technique, which has gradually gained wider acceptance for clinical application. It has a high detection rate when used to examine nerves thickening, oedema, lesions discontinuity with adjoining soft tissue, and tumours. In general, high-frequency ultrasound is...
able to provide high-quality sonogram images of the brachial plexus\cite{13} and is fast gaining popularity\cite{14}. (e) Other imaging techniques such as positron emission tomography (PET) could also be used to assist the clinical staging of tumours and to monitor their condition in the affected bones and signs of distant metastasis. Apart from this, the efficiency of imaging-assisted biopsies could also be improved as research has shown that biopsies that are performed under the guidance of CT have a higher positive rate than using fibre-optic bronchoscopy\cite{8}. (3) In laboratory tests, the levels of tumour markers have also been shown to be helpful in providing a correct diagnosis of the disease.

Treatment: Currently, this disease has no dedicated treatment protocol as demonstrated by our case study. Patients can only rely on radiation therapy, chemotherapy, or other non-surgical treatment in the absence of viable surgical procedures. Symptomatic treatment mainly involves controlling a patient’s nutrition and addressing the pain experienced. Properly administered morphine is not addictive, and generally improves the patient’s quality of life while nerve block therapy is only a temporary treatment. For those who are suffering from severe pain but are unable to undergo resection, they can opt for nerve tumour dissection\cite{11}. Meanwhile, for those who have been diagnosed with superior pulmonary sulcus tumour, they could consider surgical treatment. Mediastinoscopy examination could be performed before the surgery to establish an accurate staging of the disease\cite{15}; however, the complete resection rate (50%) of superior pulmonary sulcus tumour and the 5-year survival rate basically remain unchanged. Radiation and chemotherapies could be applied to relieve the symptoms of patients suffering from metastasis. Even though a comprehensive treatment protocol significantly increases the long term survival rate of patients, the overall prognosis is still relatively poor after undergoing complete resection, with a 30% 5-year survival rate for patients receiving adjuvant radiation therapy\cite{12}. Affected patients are mostly in suffering due to disease progression, and subsequently die of cachexia or respiratory failure.

Through the treatment of this patient, we have gained some vital understanding of the disease. (1) The patient had a previous history of lung cancer and had undergone surgery. He experienced pain that gradually spread through his arm, along with the manifestation of ipsilateral Horner syndrome; his condition was eventually diagnosed as Pancoast syndrome. However, it remains unknown whether his condition was caused by the trauma of an apical primary tumour or clavicular lymphadenectomy. Therefore, it is important to be aware of the presence of inflammation when diagnosing this disease. (2) Kenji’s retrospective studies have shown that Pancoast syndrome has a higher rate of misdiagnosis\cite{14}. Two-thirds of doctors are unfamiliar with this disease, of which 29% of them are orthopaedists involved in giving the initial diagnosis, and only 25% of patients have a complete set of imaging examination data. (3) B-scan ultrasonography showed serious injury in the lower trunk of brachial plexus, which formed traumatic neuroma. This shows that B-scan ultrasonography is useful in the diagnosis of this disease. (4) The patient experienced severe pain without sustaining any injury. With all the aforementioned characteristics, we believe that the patient’s first lymphadenectomy surgical experience affected the disease. As such, neurolysis was performed on the brachial plexus. During surgery, the anterior scalene was found to be heavily scarred with a changed appearance, and the lower trunk of brachial plexus was strongly adhered to the surrounding tissue. After neurolysis, the patient’s pain was significantly relieved. We believe that neurolysis is not a common treatment and not every patient will benefit from it. As for the treatment of superior pulmonary sulcus primary tumour, tumour resection is the best solution because an unnecessary neurolysis is evidently risky.

In short, the cause of Pancoast syndrome is complex and involves a variety of reasons. It does not appear to have specific symptoms in the early stages, thus complicating diagnosis. As such, it is important for medical practitioners to gain a better understanding of the clinical manifestations of Pancoast syndrome to improve a patient’s prognosis.

Conflict of interest

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

References

5. Sang J, Gong Z, Xu X. Diagnosis and treatment of lung