Rare disease

Shoulder pain and isolated brachial plexopathy

Amar U Kishan,¹ Sana Syed,² Franchesca Fiorito-Torres,² Manisha Thakore-James^{2,3}¹Department of Health Sciences and Technology, Harvard Medical School, Boston, Massachusetts, USA²Department of Neurology, Boston University, Boston, Massachusetts, USA³Department of Neurology, Boston VA Healthcare, Boston, Massachusetts, USA

Correspondence to Dr Amar U Kishan, aukishan@gmail.com

Summary

Pancoast syndrome, classically considered as a constellation of (1) pain along the C8–T2 dermatomes, (2) weakness and atrophy of the hand and (3) Horner's syndrome, often presents a diagnostic challenge. In fact, it may manifest as a singular orthopaedic complaint, prompting a futile barrage of tests and referrals. The authors present the case of an elderly man who initially presented with severe shoulder pain. Due to progressive pain and weakness, he was referred to rheumatology and was treated with corticosteroid injections for a presumed musculoskeletal lesion. Ultimately, he manifested gross muscular atrophy and worsening pain, prompting a referral to neurology. An electromyogram (EMG) suggested a lower brachial plexopathy, and a follow-up brachial plexus MRI identified a large Pancoast tumour. Unfortunately, his disease was rapidly progressive, and he passed away within 2 months. While the MRI remains the gold standard for diagnosing Pancoast syndrome, an EMG can facilitate diagnosis in difficult cases such as this one.

BACKGROUND

Pancoast syndrome often presents a diagnostic challenge due to its variable presentation. The classic constellation of pain along the C8–T2 dermatomes, weakness and atrophy of the hand, and Horner's syndrome is an uncommon manifestation. Instead, patients generally complain of isolated symptoms of shoulder or limb pain, which can often lead to an extensive battery of futile diagnostic and therapeutic procedures. This is an illustrative case that underscores the importance of considering Pancoast syndrome in such instances and highlights the diagnostic utility of an EMG in facilitating accurate diagnosis.

CASE PRESENTATION

A 70-year-old right-handed man was referred to the neuromuscular clinic for evaluation of 2 months of pain and atrophy of the left (L) medial forearm. Nine months prior, he was evaluated by his primary care physician for new L shoulder pain. At that time, his examination and plain films of the shoulder were unremarkable. He was referred to a rheumatologist and diagnosed with atypical myofascial pain syndrome, but did not improve with corticosteroid injections. He then had an MRI of his L shoulder, which showed tears in the supraspinatus muscle and infraspinatus tendon, and began a physical therapy programme. At a subsequent follow-up visit, he complained of worsening of his pain and was noted to have muscular atrophy of the L thenar and first dorsal interosseus muscles, prompting referral to the neuromuscular clinic.

At the clinic, he described the pain as a constant, sharp, 10/10 L suprascapular pain radiating to his L medial forearm and palm, with numbness in the L fourth and fifth digits. The pain was mildly relieved by rest and greatly aggravated by movements of the upper extremity, leading to significant difficulty in completing activities of daily living. High-dose non-steroidal anti-inflammatory drugs and topical capsaicin had no effect on the pain. On

review of systems, he complained of 14 pounds of weight loss over the past 6 weeks, fatigue and difficulty in sleeping due to pain, but denied fever, chills, night sweats, cough, shortness of breath and hoarseness. His social history was significant for a 50-pack-year history of cigarette use, while his family history was non-contributory.

He worked as a porter but could not recall any inciting trauma. Supporting laboratory results included HbA1c 6.1, erythrocyte sedimentation rate (ESR) 16 and CK 59.

On examination, he was noted to be frail and in significant discomfort. His motor examination was limited proximally due to severe pain. His motor examination was notable for fasciculations of the L flexor carpi ulnaris and marked atrophy of the L thenar, hypothenar, interossei muscles and flexor carpi ulnaris muscles, and decreased strength distally greater than proximally on the L. Specifically, he had 3/5 strength in the L first dorsal interosseus, abductor digiti minimi (ADM) (ulnar, C8 and T1) and in the L anterior interosseous distribution (flexor digitorum profundus (digit 2) and flexor pollicis longus) and could not form an 'ok' sign. He had 3/5 strength in the finger flexors, triceps brachii, wrist flexors, wrist extensors, pronator teres, opponens pollicis, abductor pollicis brevis and finger extensors. His sensory examination was notable for decreased sensation to light touch and pinprick in the medial R forearm with hyperaesthesia of his palm and fourth and fifth digits. Vibratory sensation was normal. Deep tendon reflexes were 1+ in the L upper extremity, but were otherwise 2+ throughout. Hoffman's and Babinski's signs were negative. The remainder of his examination was unremarkable.

INVESTIGATIONS

An EMG and nerve-conduction-study (NCS) were obtained to further characterise the weakness and atrophy. The EMG/NCS revealed complicated findings (table 1). Motor conduction studies showed reduced

Table 1 Nerve conduction studies and electromyogram

Motor (CMAP)	Distal latency (ms)		Distal amplitude (mV)		Conduction velocity (m/s)
Left median-APB	4.60 (prolonged)		1.1 m (reduced)		45.3 (slow)
Right median-APB	3.9		6.5		53.4
Left Ulnar-ADM	3.35		6.7		56.1
Right Ulnar-ADM	3.2		10.2		58.8
Sensory (SNAP)	Peak onset (ms)		Distal amplitude (uV)		Conduction velocity (m/s)
L Median	3.2		20.0		49.1
R Ulnar	3.0		13.4		44.9 (reduced)
L Ulnar	Absent		Absent		Absent
Muscle	Insertional activity	Fibrillation potential	Motor unit amplitude	Motor unit duration	Motor unit recruitment
L pronator teres	Mod increase	Severe	Mild increase	NI	Reduced
L FDI	Mod increase	Severe	Mod increase	NI	Reduced
L biceps	NI	NI	NI	Mild increase	NI
L triceps	NI	NI	NI	Mod increase	Reduced

CMAP, compound muscle action potential; SNAP, sensory nerve action potentials.

median motor compound muscle action potential (CMAP) amplitudes indicating axonal pathology of the L median nerve to the abductor pollicis brevis (APB; C8–T1 roots). He was also noted to have evidence of reduced ulnar motor CMAP amplitude, compared with the unaffected right side, indicating mild axonal pathology of the ulnar nerve (ADM, C8–T1 roots). The L ulnar sensory

Figure 1 MRI of the left brachial plexus showing a large superior sulcus tumour.

nerve action potentials (SNAP) is absent (C8 sensory distribution). However, the L median SNAP was normal (C6 sensory distribution). These are the classical nerve conduction findings of a lower trunk of the brachial plexus lesion. The medial antebrachial cutaneous sensory nerve will also be affected in this condition, but was not tested in this patient due to his difficulty with tolerating the test. The EMG was very limited in our patient because the test was very painful for him. This part of the study showed evidence of active, ongoing denervation injury in the L pronator teres (C7 myotome) and first dorsal interosseus (C8, T1 and myotomes). None of the other muscles tested showed these acute changes.

The gravity of the possible diagnoses and suboptimal pain control prompted an admission to the neurology inpatient service and a chest x-ray, rheumatological panel and brachial plexus MRI were ordered. The chest x-ray was normal, but the MRI revealed a L upper lobe 3.7×3.5×4.2 cm superior sulcus tumour extending into the middle and lower trunks, T1 nerve root, medial and posterior cords and L T1 rib (figure 1). The tumour encased the L subclavian artery after the takeoff the vertebral artery. A repeat ESR and C-reactive protein (CRP) were elevated 34 and 35, respectively, but the rheumatological workup (ANCA, ANA, RF, C3 and C4) and serum electrophoresis were negative.

DIFFERENTIAL DIAGNOSIS

It was difficult to clinically correlate the results of these studies, but the examination suggested that the majority of the symptoms were secondary to a L lower brachial plexopathy involving the C7–T1 nerve roots. However, depressed deep tendon reflexes throughout the L upper extremity and the slight biceps brachii weakness indicated that a more diffuse brachial plexopathy could be responsible, with clinical involvement being more marked in the lower trunk. Unfortunately, the patient's inability to tolerate an extended examination precluded further analysis by EMG/NCS. Regardless, the limited neurophysiological studies suggested a diffuse brachial plexopathy, rather than a simple orthopaedic complaint. The severity of his pain supported an inflammatory or neoplastic process, such as Parsonage-Turner syndrome or idiopathic brachial plexopathy. Given his weight loss and history of smoking, a specific concern was a superior sulcus tumour causing Pancoast syndrome. Finally, mononeuritis multiplex was another possibility. While the rotator cuff injury identified on MRI could have contributed to his shoulder pain, the neurological symptoms, degree of atrophy and progression of his symptoms despite appropriate treatment implied an alternative disease process.

OUTCOME AND FOLLOW-UP

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The patient was transferred to the Internal Medicine service for further diagnostic workup and discussion on therapeutic goals. A CT scan of the chest, abdomen and pelvis confirmed a spiculated mass at the L lung apex with mild paratracheal lymphadenopathy but no clear evidence of metastatic disease. A brain MRI showed no evidence of intracranial metastases. A bronchoalveolar lavage found no malignant cells, so a CT-guided biopsy was performed. The biopsy showed a poorly differentiated non-small-cell lung carcinoma (NSCLC), and he was staged as having American Joint Committee on Cancer (AJCC) stage IIIA T4N0M0 disease. His pain medication regimen at discharge included MS contin 15 mg twice daily with oxycodone 10 mg every 4 h for breakthrough pain, 300 mg gabapentin three times a day and dexamethasone 4 mg twice daily for neuropathic and inflammatory components of his pain. He was discharged to a rehabilitation facility to undergo intensive physical therapy and will receive epidural corticosteroid injections for pain control with neoadjuvant chemoradiotherapy. Unfortunately, he succumbed to his disease within 2 months.

DISCUSSION

Pancoast syndrome has many inflammatory and neoplastic causes and the classic presentation is pain along the C8–T2 dermatomes, weakness and atrophy of the intrinsic muscles of the hand and Horner's syndrome (ptosis, miosis and anhidrosis).¹ The eponymous Henry Pancoast published the first exhaustive case series describing this syndrome and its radiological correlates in 1924,² concluding that the tumorigenic tissue was epithelial tissue from the fifth branchial cleft. Pancoast later published a comprehensive expansion of the original series in 1932³; unbeknownst to him, José Tobías simultaneously described the syndrome and correctly surmised the site of origin of the tumour as bronchopulmonary tissue.⁴ These tumours are mostly NSCLC, with adenocarcinomas slightly more common than squamous carcinomas. Pancoast tumours are rare, and constitute less than 5% of NSCLCs.⁵ Notably, a wide variety of other entities can also cause Pancoast syndrome, including aspergillosis, tuberculosis and lymphoma.¹

In this patient, the diagnosis of Pancoast syndrome was very likely at the time of initial consultation, given the patient's intense shoulder pain, history of smoking, weight loss and subsequent neurophysiological evidence of lower brachial plexopathy. However, this case presents several worthwhile points for consideration. First, while Horner's syndrome is classically considered a hallmark of Pancoast syndrome, it should be remembered that its prevalence in published case series is quite variable, ranging from 4% to 64%. Conversely, shoulder pain and arm pain or 'brachialgia' can be relatively common presenting complaints, ranging from 19% to 65% in the same series. As such, many patients have exhaustive rheumatological and orthopaedic evaluation, leading to delays in diagnosis of up to 7.5 months.^{6–8} In the case of our patient, the diagnosis was complicated by his underlying orthopaedic history of shoulder bursitis and the rotator

cuff tears on MRI of the shoulder. Red flags for a more sinister process included the lack of an inciting trauma, the progressive nature of the pain and muscular atrophy and the persistence of pain despite numerous interventions. In retrospect, it is highly likely that part of his shoulder pain was due to referred irritation of the parietal pleura, and that the subsequent brachial plexopathy merely represented progression of the Pancoast tumour, instead of being its true original presentation.

MRI remains the imaging modality of choice for diagnosing a superior sulcus tumour, and plain films in particular are quite poor in making the diagnosis, as was the case in this patient.¹ Data on the diagnostic utility of EMG/NCS in Pancoast syndrome are less exhaustive. A study comparing findings between 35 patients with neoplastic or radiation-induced plexopathy found that the distribution of weakness on NCS was of little diagnostic utility, while the most useful discriminating EMG finding was the presence of myokymic discharges in the case of radiation plexopathy.⁹ A case of isolated cervical radiculopathy with normal sensory studies secondary to Pancoast tumour, diagnosed by EMG, has been reported.¹⁰ However, for the aforementioned reasons, the decision to obtain an MRI or neurophysiological study appears to be the rate-limiting factor in diagnosing Pancoast syndrome. In this patient, the EMG/NCS studies showed abnormality of the L median motor (C8–T1) and absence of the L ulnar sensory potential (C7) with sparing of the L median sensory potential (C6) which is characteristic of a lower brachial plexopathy. The EMG/NCS findings coupled with the patient's clinical symptoms provided clear evidence of a lower brachial plexopathy. In addition, his associated symptoms of weight loss and history of smoking with the above clinical picture and test results greatly increased the post-test probability of our patient having a Pancoast tumour. If the patient had been able to tolerate a more extensive EMG/NCS, it would have been helpful in further delineating the extent of the brachial plexopathy. Nonetheless, even with an abridged EMG/NCS, we believe that there is a possible ancillary role for neurophysiological testing in directing the course of diagnosis in challenging cases of Pancoast syndrome.

Learning points

- ▶ Making the diagnosis of Pancoast syndrome can be challenging when the patient presents with isolated, vague complaints. Unfortunately, this is often the case, since the 'classic' presentation is rare.
- ▶ An MRI of the brachial plexus remains the gold standard diagnostic test, but this detailed study is often used as a confirmatory procedure once pretest probability is high.
- ▶ In difficult scenarios, an electromyogram can be a useful ancillary tool in narrowing the differential diagnosis when faced with vague presentations.

Competing interest None.

Patient consent Obtained.

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