"My arm is on fire!"

DALHOUSIE UNIVERSITY Inspiring Minds

Rose Mengual, MD, ACP

John's case

John is a 49-year-old male who presents to the ED with complaints of right upper back and shoulder pain worsening over 6 months.

John describes his pain as "sharp and burning" with radiation down the medial arm, forearm and third to fourth digits. The pain is accompanied by intermittent numbness alternating with allodynia along the same distribution as pain. Over the last 2 months, he notes decreased dexterity in his right hand. He describes nocturnal pain that has become so severe he is sleeping on the floor without a pillow or mattress. Lastly, he describes having a "lazy eye" on the right side for the last 2 months. John denies fever, IV drug use, or any inciting trauma.

He was seen in the ED 2 months prior where he underwent plain film radiography of the cervical and thoracic spine. The plain film of the thoracic spine is reported as normal and cervical spine as demonstrating mild disc space narrowing at C5-C6 and C6-C7 with osteophytic narrowing of the C5-C6 nerve root foramina on the right. He was diagnosed with osteoarthritis of the cervical spine and discharged with a prescription for acetaminophen with codeine and follow-up by his FP.

John's FP has arranged a follow-up CT spine and placed a referral for outpatient neurology consultation.

Read on for more on John.

Questions & Answers

1. What is the differential diagnosis of neuropathic pain?

Neuropathic pain may originate from various processes that affect the brain, spinal cord, or peripheral nerves. Therefore, it is useful to consider neuropathic pain as having a central or peripheral etiology. The differential diagnosis is extensive (Table 1) but should focus initially on ruling out life-threatening or treatable conditions.

2. What are the most likely anatomic localization of John's syndrome?

The presence of neuropathic pain in a dermatomal pattern (C7, C8 and T1) as well as loss of power in the hand suggests a proximal process affecting both afferent sensory tracts as well as efferent motor pathways to the entire limb. This suggests that the most likely anatomic localization of John's pathology is the spinal cord (*i.e.*, cord compression affecting both ascending sensory tracts and descending corticospinal tracts), nerve roots (*e.g.*, compression at the foramina), or the brachial plexus (*e.g.*, invasion or external compression) where both afferent sensory and efferent motor pathways remain paired. The most likely diagnoses in John's case include a compressive myelopathy, nerve compression or infiltration by tumour, or severe and progressive radiculopathy.

3. How should John be investigated?

Pathology affecting the spinal cord, nerve roots, or brachial plexus may arise from the spinal cord, spinal column, surrounding soft tissues, or extension from adjacent

Table 1
Differential diagnosis of neuropathic pain

Central	Peripheral
Compressive myelopathy from spinal stenosis - Disc herniation - Epidural abscess - Malignancy	Radiculopathy - Cervical - Thoracic - Lumbar
Multiple sclerosis	Nerve compression or infiltration by tumour
Post-ischemic myelopathy	Entrapment neuropathy
Post-radiation myelopathy	Postherpetic neuralgia
Post-stroke pain	Post-traumatic neuralgia
Post-traumatic spinal cord injury pain	Nutritional deficiency-related neuropathy
HIV sensory neuropathy	Alcoholic polyneuropathy

John's case cont'd

Medical history

- Cigarette smoking: 44 packs per year
- Carpal tunnel release
- Cholecystectomy
- Vitamin B_{12} deficiency

Medications

- Acetaminophen 500 mg with codeine 30 mg two tabs p.o. q.4.h.
- Vitamin B_{12} injection once monthly

Examination

- Temperature: 36.3ºC
- BP: 138/60 mmHg
- Heart rate: 84 bpm
- Respiratory rate: 16 breaths per minute
- Oxygen saturation: 97% on room air

On examination, John is alert and oriented to person, place and time. He is sitting upright and occasionally groans with movement. He has right-sided ptosis and miosis. His cervical spine is non-tender to palpation and he has normal neck range of motion. He has diffuse tenderness of the right upper back superior to the scapular spine. He has allodynia and decreased sensation to light touch of the right medial arm, forearm and third to fifth digits. Power is 5/5 in all myotomes of the upper extremity with the exception of digit abduction and wrist extension where power is noted to be 4/5. His right triceps reflex is absent. His complete blood count and electrolytes are within normal limits.

Radiology (Figures 1a, 1b and 2)

Read on for John's conclusion.

body cavities including the chest. Plain film radiography of the cervical spine and chest are the most appropriate initial investigations. Given the severity, duration and progressive nature of John's condition, an urgent referral for neurologic or neurosurgical evaluation with targeted imaging of the spine, spinal cord, nerve roots and brachial plexus by MRI with or without preceding CT scan would be warranted in the absence of a clear etiology on plain film radiography.

4. *What is Pancoast's syndrome?*

Pancoast's syndrome is a collection of signs and symptoms most commonly resulting from extension of an apical lung tumour at the superior thoracic inlet. Invasion of brachial plexus, pleura, vertebrae and soft tissues leads to classic findings of shoulder, chest and/or upper extremity pain, hand weakness and Horner's syndrome. A delay to diagnosis of up to 10 months is common as many patients are often initially diagnosed with and treated for shoulder bursitis or cervical spine osteoarthritis.

Invasion of the C7 to T1 nerve roots results in paresthesia, allodynia, numbness or neuropathic pain of the medial arm, forearm and third to fifth digits. Invasion of these nerve roots also accounts for weakness and atrophy of the intrinsic hand muscles.

Horner's syndrome is a constellation of clinical findings resulting from the invasion of the cervical paravertebral sympathetic chain (stellate ganglion) and is observed in 20% to 50% of patients with Pancoast's syndrome. Findings of Horner's syndrome include ipsilateral miosis, ptosis and anhydrosis.

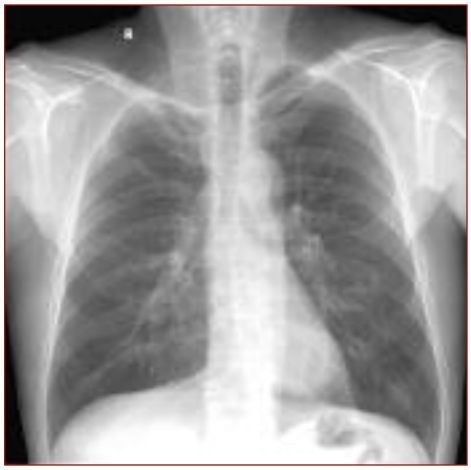

Figure 1a. Posteroanterior chest x-ray. Apical pleural thickening on the right suggestive of a superior sulcus or Pancoast's tumour with bone destruction of the medial portion of the right second rib.

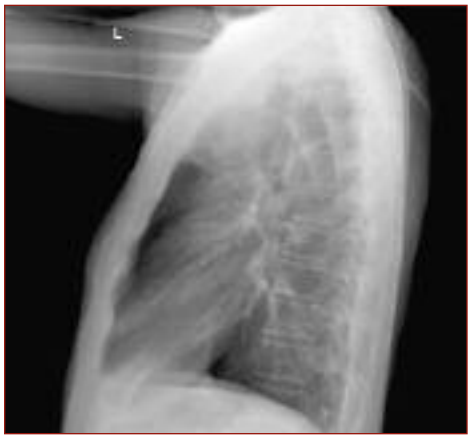

Figure 1b. Lateral chest x-ray. Possible lymphadenopathy.

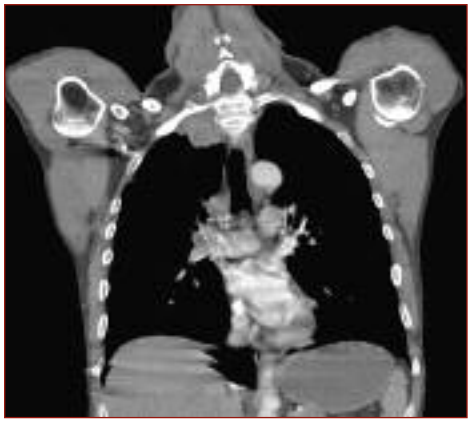

Figure 2. CT scan. A superior sulcus or Pancoast's tumour in the right apex.

5. What is the etiology of Pancoast's syndrome?

Pancoast's syndrome may result from a vast array of pathologic processes of neoplastic, infectious, or inflammatory etiology. By far, the most common etiology is non-small-cell primary bronchogenic carcinoma. The remaining differential diagnosis is broad and includes other primary thoracic neoplasms (*e.g.*, mesothelioma), metastatic neoplasms (*e.g.*, thyroid, larynx, bladder), hematologic neoplasm (*e.g.*, lymphoma) and infectious conditions.

6. What findings on chest x-ray suggest the presence of a Pancoast's tumour?

The following findings on chest x-ray are suggestive of a Pancoast's tumour:

- Unilateral apical cap > 5 mm
- Asymmetry of bilateral apical caps > 5 mm
- Bone destruction
- Apical mass

Pancoast's tumours can also be detected on lower cervical spine and upper thoracic spine plain film radiographs.

7. What confirmatory testing is needed?

An abnormal plain film chest radiograph suggestive of Pancoast's tumour should be followed-up with enhanced CT scan of the chest. A cytologic diagnosis is most successfully obtained by percutaneous transthoracic CT guided biopsy.

8. What is John's prognosis?

The mainstay of treatment for Pancoast's tumour is preoperative radiation therapy followed by extensive surgical resection unless there is evidence of extensive invasion (*e.g.*, brachial plexus, base of neck) or distant spread. For tumours not amenable to surgical resection, the mainstay of treatment is radiation therapy with or without chemotherapy. Five-year survival rates range from 0% to

35% depending on disease burden and treatment options available at the time of diagnosis.

9. *What are the treatment options for John's neuropathic pain?*

Successful palliation of neuropathic pain requires an individualized approach for each patient. Treatment with acetaminophen and/or NSAIDs is often ineffective in management of neuropathic pain. Guidelines for the management of neuropathic pain[1] suggest first-line treatment with gabapentin, a tricyclic antidepressant, or a selective serotonin-norepinephrine reuptake inhibitor while taking into consideration the patient's medical comorbidities and medication side-effects. These agents may be supplemented by the addition of tramadol or an opioid analgesic. Dx

John's case cont'd

John is referred to the Thoracic Surgery service for admission, analgesia and further investigation. He is started on hydromorphone and gabapentin for management of his neuropathic pain.

In hospital, John underwent a percutaneous transthoracic CT guided biopsy which confirmed the presence of primary squamous cell carcinoma of the right lung. Given the presence of adrenal metastases and degree of vascular and brachial plexus involvement, surgical resection was not a treatment option. John was started on palliative chemotherapy and radiation therapy on an outpatient basis.

Dr. Mengual is a Fourth Year RCPSC Emergency Medicine Resident, Dalhousie University and Queen Elizabeth II Health Sciences Centre, Halifax, Nova Scotia.

Reference
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Resource
1. Arcasoy SM, Jett JR: Superior Pulmonary Sulcus Tumors and Pancoast's Syndrome. N Engl J Med 1997; 337(19):1370-6.

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