

AN 80-YEAR-OLD WOMAN WITH LEFT SHOULDER PAIN

SB Khoo FRACGP, Penang Medical College

Address for correspondence: Dr Khoo Siew Beng, Senior Lecturer & Family Physician, Penang Medical College, No 4, Sepoy Lines Road, 10450 Penang, Malaysia. Tel: 604-2263 459, Email: siewbeng@gmail.com

ABSTRACT

This case history illustrates the real life experience and dilemma of an 80-year-old woman in pursuit of medical care for her left shoulder pain. Points for discussion range from clinical features of Pancoast tumor, importance of pain management, good principles of Family Medicine and Palliative care to ethical issues of conspiracy of silence, limited treatment plan and palliative versus curative radiotherapy treatment without a known biopsy report. This paper provides opportunity for analysis of a real complex clinical situation, application of medical knowledge to problem solving in clinical practice and relevant topics for discussions. (For anonymity sake, the names of patient, doctors, general and private hospitals are not mentioned. The aim of this paper is solely for continuous medical education without any intention to ridicule any party).

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CASE HISTORY

Madam YN was an 80-year-old Chinese lady who presented with pain in her left shoulder for two months since the end of year 2007. Her daughter took her to see private general practitioners (GPs) who prescribed her some painkillers with the reassurance that it was "rheumatism due to old age" that should improve with rest.

At the initial stage, the pain was confined to the inferolateral aspect of the left scapula. Her pain became worse and gradually extended down her left arm and forearm at the medial ulnar aspects. She described the pain as continuous "deep, drilling and gnawing" at the shoulder and excruciating, episodic "cutting, lancinating and lightning type of pain" down the left arm. Intensity of the pain was variable. Initially there was temporary relief with massages, topical salicylate ointment and oral painkillers but towards the end the pain became unbearable. During the period between the onset of pain at the end of year 2007 and August 2008, she had consulted more than ten different private GPs and hospital specialists with overall 20-30 consultations including a GP pain specialist who advised her to go for acupuncture. Medications prescribed for her ranged from oral paracetamol to non-steroidal anti-inflammatory drugs (NSAID), tramadol and DF 118 (mild opioids) to transdermal ketoprofen. X-rays were done twice in a private hospital A; there was no fracture detected and YN was referred for physiotherapy. YN requested for hospital admission in August 2008 because she could no longer tolerate the pain but she was told that this was due to old age, that there was no specific treatment for it to warrant hospitalization.

Madam YN's medical history was unremarkable except for diabetes mellitus that was well controlled with oral gliclazide

and metformin. Other than moderate bilateral hearing impairment, she was generally well physically and mentally, was independent and with good assessment scores for activities of daily living.

In May 2008, YN noticed her left eye had become smaller and there was drooping of the upper eyelid on the same eye. She consulted an ophthalmologist who screened her for diabetic retinopathy and informed her there was nothing to worry about.

In November 2008, YN's daughter decided to take her to see a neuro surgeon "friend" in a private hospital B who did another left shoulder X-ray. Incidentally, he discovered a mass at the apex of the left lung. CT scan of the thorax confirmed the presence of a tumour at the apex of the left lung, about 5 cm in diameter with irregular borders. YN was referred to a general physician as there was no chest physician in that private hospital. A review of the previous X-ray done three months earlier at private hospital A showed that the mass was already there. It was slightly smaller than the current size. It was missed by the radiologist who reported as "No fracture or dislocation seen". In view of YN's age the physician advised that YN should not be subjected to further biopsy tests in pursuit of curative treatment but only to relieve her pain with low dose palliative radiotherapy.

YN's children requested that their mother must not be told of the diagnosis; she would not be able to take it.

YN was entitled to free treatment at the General Hospital (GH) as her children were in the government service. She was referred to Oncology Clinic of a Public General Hospital. Needle biopsy of the tumour mass conducted under ultrasound guidance was attempted twice, but no malignant cells were

seen on both occasions. In view of the negative results, GH oncologist could not prescribe radiotherapy treatment but prescribed aqueous morphine 10 mg *tds.* for her pain instead.

After taking two doses of 10 mg morphine for the first time, YN became drowsy, nauseated and slept the whole day. She developed delirium. She was talking incoherently, could not recognise her children, refused to eat, and was seeing visual images of relatives who had passed away years ago. Her blood pressure increased to 180/100 mm Hg, glucometer recorded a random blood sugar of 3.5 mmol/L. Frantically her daughter took her back to private hospital B for admission. Head CT scan done did not show any brain metastases, MRI of spine did not show vertebral infiltration of malignant cells or spinal cord compression and her serum calcium was normal. After two days of showing no improvement, YN's daughter requested for her mother to be transferred to a private Oncology hospital C where radiotherapy facilities were available.

Low dose palliative radiotherapy (DXT) was delivered with the intention to just control the pain. However after delivering five fractions of DXT, the tumour mass was found to have shrunk considerably. The radiotherapist decided to carry on with DXT treatment to a total course of 25 fractions. YN's delirium did not seem to improve but she was more comfortable where pain was concerned.

YN's delirium cleared the day after coming home. Her family decided not to let her go back for hospital readmission. She continued her radiotherapy treatment as a day care patient till completion of the full course.

YN never recovered from her inability to walk. She gradually developed loss of sensation, paralysis and wasting of muscles of left hand, pleural effusion, progressive hoarseness and loss of voice. Pain was under control but she continued to lose weight, became more anaemic and anorexic and her general

Table 1

Key features for discussion	
Encounters	Related topics for discussion
<p>1. Difficulties in diagnosing Pancoast tumour</p> <p>a. Why did it take so long to diagnosis Pancoast tumour in spite of YN's consultations with so many GPs, public and private hospital specialists?</p> <p>b. Why was YN's request for hospitalization turned down at private Hospital A? Why did the ophthalmologist miss the diagnosis of Horner's syndrome? Why did the radiologist miss the tumour mass in the apex of the left lung in August 2009? Why were YN's pain and eye problem not detected by the endocrinologist at her three monthly review for diabetes?</p>	<p>Non typical presentation of Pancoast tumour. Inadequate pain assessment. "Doctor hopping and shopping". Healthy aging process. Chronological age versus functional age. Missing cues. Premature convergence of hypothesis. Practice of "compartmental medicine".</p>
<p>2. Pain management</p> <p>a. Why must YN endure the pain for so long?</p> <p>b. Was it ethical to call off all painkillers and focused on control of hypertension and diabetes?</p>	<p>Inadequate pain management. Collusion of anonymity. Prioritization for pain control in palliative care.</p>
<p>3. Ethical issues and limited treatment plan</p> <p>a. Was it ethical to forego further investigations and curative treatment based alone on patient's age?</p> <p>b. Was it ethical to subject YN to radiotherapy treatment if there was no positive HPE report of malignancy?</p> <p>c. Was it ethical in hiding the truth from YN?</p>	<p>Curative versus palliative radiotherapy. Limited treatment plan/patient autonomy. Risk versus benefits of low dose palliative radiotherapy for pain control without HPE results. Disadvantages of conspiracy of silence.</p>
<p>4. Palliative care</p> <p>a. Why did YN behave abnormally after taking the morphine?</p> <p>b. Were total bed confinement, IV infusion, Ryle's tube feeding and catheterization in-situ necessary?</p>	<p>Contributing factors to patient's delirium. Acute hospital care versus home palliative care for end-of-life cases. Definition of a good death.</p>

condition progressively deteriorated. She was going in and out of hospitals B and C for symptom management and was confined to bed till the end. She was admitted into private hospital B for five days before she passed away in the same hospital at the end of September 2009.

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DISCUSSION

This case history illustrates the dilemma of an elderly woman with left shoulder pain, in her search for a correct diagnosis, appropriate investigations, specific treatment and palliative care. Key features for discussion in this case are determined by the events encountered by the patient. Table 1 gives a summary of these encounters under appropriate headings with related topics for discussion.

1. Difficulties in diagnosing Pancoast tumour

Non-typical presentation of Pancoast tumour

Pancoast tumour is a malignant tumour of the superior sulcus of the lung. Although this is a lung tumour, pulmonary symptoms of cough, dyspnoea and haemoptysis are rare. Very often by the time it is discovered, it has infiltrated to the surrounding brachial plexus and cervical sympathetic trunk.¹ Pancoast syndrome is the term given to a combination of constant and characteristic clinical presentations of pancoast tumour.^{1,2}

1. Severe pain localised in the shoulder (90% of cases) - Pain may radiate towards the axilla and scapula and ulnar aspect of the upper limb along C8, T1 dermatomal distribution with or without atrophy of the arm and hand muscles.
2. Horner's syndrome (62% of cases) - Involvement of sympathetic chain results in ptosis, miosis, hemianhydrosis and enophthalmus.
3. Invasion or destruction of ribs and vertebrae.
4. Superior vena cava syndrome (not a typical presentation of Pancoast syndrome).

In the early stages, Pancoast tumors are difficult to detect on chest X-ray films because the top of the lung is located in an area of the body that is difficult to visualize clearly on an X-ray film. Many patients are initially treated for presumed local musculoskeletal problems of bursitis and vertebral osteoarthritis with radicular pain.^{1,2} In 1998 series by Maggi

and colleagues, symptoms of Pancoast tumour was shown to last for 2-36 months with a mean duration of 9.7 months.³

Healthy aging process/missing cues/prematured convergence of hypothesis of doctors/compartmental medicine/ "doctor hopping and shopping" of patient.

Normal aging refers to the common complex of diseases and impairments that characterise many of the elderly. In successful healthy aging deleterious effects are minimized, functions are preserved, and aging is not necessarily accompanied by debilitating disease and disability.⁴

Chronological age refers to the number of years a person has lived. Functional age refers to a person's ability to contribute to society and benefit others and himself. Many people may be chronologically old but remain physically fit, mentally active and productive members. Comprehensive health assessment, the cornerstone of quality care for the elderly, is a multidimensional assessment that incorporates physical, psychological and social function as well as medical health.⁵ There was no justification to assume that because YN was already 80 years old, there was nothing more to be done for her "rheumatic pain".

Presence of a mass in the apex of the left lung was obvious on the X-ray film done in August 2008 but was not picked up by the radiologist. As neuropathic pain was not identified, doctors focused their attention on nociceptive pain due to arthritis or possible bone fracture even though there was no history of a fall. The radiologist confined his attention to looking for fractures on the X-ray film as ordered by the physician, so much so that he missed out the big mass at the apex of the left lung. For the same reason, the endocrinologist and ophthalmologist missed out the presence of neuropathic pain and Horner's syndrome because they focused only on diabetes mellitus and diabetic retinopathy. Premature convergence of hypothesis led to failure to 'search for further hypothesis, missing other more important facets of the whole'.⁶ To avoid making this mistake doctors should try to include all appropriate diagnostic hypotheses based on probability, seriousness, 'pay off' and treatability.⁷

Presence of Horner's syndrome associated with radicular pain should have led to the suspicion of an infiltrative lesion at the thoracic outlet involving the brachial plexus and extending to the sympathetic chain. Medical sub-specialization has led to practice of "compartmental medicine" where each specialist looks after a particular organ system. The many doctors and specialists consulted by YN failed to relate the eye problem to the shoulder pain, combination of which gave a strong clue to the diagnosis of Pancoast syndrome.

"Doctor hopping and shopping" by patient is common practice in our Malaysian healthcare system. No one particular doctor

was responsible or committed to looking after YN's shoulder pain. Michael Balint in his concept of "collusion of anonymity" emphasized that "if a patient is seen by a variety of physicians, not one of them is willing to be responsible or accept the total management of the patient's problems".⁸

2. Pain management

Pain assessment

Pain is one of the more easily controlled symptoms in palliative medicine. Cancer pain can be effectively treated in 85% to 95% of patients with an integrated program of systemic, pharmacologic and anticancer therapy.⁹

Acute pain is of sudden onset, has a clear cause, lasts for hours to days, and disappears once the underlying cause is treated. It is beneficial to the patient as it signals that there is something wrong and motivates the person to get help. Chronic pain usually starts as an acute pain and continues beyond the normal time expected for resolution of the problem or recurs for various other reasons. It is not therapeutically beneficial to the patient. In acute pain, attention is focused on treatment of the cause of pain whereas in chronic pain, the emphasis is laid upon reducing the pain to give relief, limit disability and improve function.¹⁰

Nociceptive pain arises from damage to tissues other than nerve fibers. It usually presents as well localized, constant, dull nagging pain or throbbing and toothache-like pain if it involves the bones. Cutaneous nociceptive somatic pain results from injury to the skin, while deep nociceptive somatic pain is from musculoskeletal tissues.¹⁰

Nociceptive visceral pain results from injury to the internal organs such as the heart, lungs, brain, stomach, intestines, liver and urinary bladder. It usually presents as diffuse, non-localized, constant, dull, aching pain or as colicky pain in hollow viscus for example abdominal and ureteric colic.¹⁰

Neuropathic pain results from damage to the nervous tissue. It can be either central when the brain and spinal cord are injured or peripheral when the peripheral nervous system is injured. Neuropathic pain is described as severe, sharp, lancinating, lightning-like, stabbing, burning, cutting, prickling, or abnormal sensations of coldness, numbness, tingling or weakness.¹¹

Inadequate pain assessment

Initially YN's pain was non specific and was confined to the shoulder, justifying the impression that it was shoulder arthritis or suspected fracture. Pancoast tumours are uncommon, comprises of only 5% of lung tumours² so it would not be the first diagnosis to come to the mind. However when localized

pain progressed to "lancinating, cutting, lightning-type" of pain extending down the medial ulnar aspect of the left arm and hand, one had to be on the alert that there was presence of neuropathic pain corresponding to dermatomal distribution of C8 and T1. This type of radicular pain is often caused by nerve root compression either in the cervical spine or on the brachial plexus. Lack of recognition of neuropathic pain further delayed the diagnosis of an infiltrative lesion in the left upper thoracic outlet.

Pain control

Neuropathic pain is widely recognized as one of the most difficult pain syndromes to treat. It presents a significant challenge to clinicians as it often does not respond to conventional analgesic therapies.¹² Adjuvant analgesics' are non-conventional painkillers with a primary indication other than pain, but with analgesic properties in some painful conditions. They include anti-epileptics (carbamazepine and gabapentin), antidepressants (particularly amitriptyline) and antiarrhythmic drugs (mexiletene). They can be effective in controlling neuropathic pain.^{13,14} Although they can be used alone, they are usually co-administered with analgesics (acetaminophen, non-steroidal anti-inflammatory drugs, opioids) when treating cancer pain.¹⁵

YN's pain was not diagnosed as nociceptive visceral pain of the left shoulder and peripheral neuropathic pain (brachial plexopathy) down the left arm for nearly a year. None of the doctors that YN had consulted prescribed adjuvant analgesics (anticonvulsant or antidepressant) for her neuropathic pain. Either her pain was not recognised as neuropathic pain or it was not known that adjuvant analgesics were helpful in relieving neuropathic pain.

3. Ethical issues

Biopsy procedures

Majority of cases of Pancoast syndrome are non-small cell lung carcinoma (NSCLC), with more than 95% located in the superior sulcus. Most of it lies outside the lung and involves the chest wall, ribs, nerve roots, lower trunks of the brachial plexus and sympathetic chain.¹ Common varieties include squamous cell carcinoma (about one third of cases), adenocarcinoma (about two third of cases) and large cell carcinoma (<10%). Small cell carcinoma is a rare cause of Pancoast syndrome (<5%).²

Non-malignant causes include lymphoma, plasma cell granuloma, infections, vascular aneurysms and cervical rib syndrome. Because of the wide variety of diseases that can produce Pancoast syndrome, a histologic diagnosis is mandatory before initiating definitive treatment. Imaging and biopsy are the cornerstones of evaluation of Pancoast tumour.²

YN's family was advised not to subject YN to further biopsy but to just refer her to a private oncologist for low dose palliative radiotherapy merely for pain control. On one hand, we could argue that it was justified not to subject patient to an invasive test for in view of her age, she would not be a likely candidate for aggressive curative treatment. Low dose radiotherapy shrinks the tumour mass and relieves the compression on the nerve mainly for pain relief.

On the other hand, without a biopsy report, was it justified to subject YN to radiotherapy? What if it was not a malignant growth? In view of the "slow growth" of the tumour, a HPE result may indicate a well differentiated radiosensitive tumour. If so, will curative doses of DXT be able to prolong survival for YN? The infiltrative nature and damaging effects of the tumour on surrounding structures suggested high likelihood of malignancy. Nevertheless, two attempts of lung biopsy were carried out but were not successful in detecting malignant cells.

Palliative versus curative DXT

In the past, superior sulcus tumours were considered inoperable and incurable because of their relative inaccessibility and extensive local invasion of the thoracic inlet. Recent clinical studies showed preoperative irradiation decreases local recurrences, prevents the growth of disseminated tumour cells, and increases survival compared with irradiation or surgery alone. The five-year survival rate after surgery is approximately 30%.¹⁶ In view of YN's age and extensive tumour infiltration, it was unlikely that surgery would be recommended.

The aim of palliative radiotherapy is to provide symptomatic relief and improve quality of life with minimal side effects. Radiation in doses of 40-60 Gy administered over a period of three weeks, elicits pain relief in 90% of patients.² In some cases, the intention of treatment between curative and palliative are not clearly distinguished. There is variation of practice among clinical oncologists.¹⁷

Without a biopsy report, the intention of delivering ten fractions of low dose radiotherapy with minimal side effects to YN merely for pain control was justified. It was later increased to 25 fractions when there was substantial reduction in the size of the tumour after delivery of the first few fractions.

Limited treatment plan

Limited treatment plan is a decision not to initiate treatment or medical interventions for the patient if benefits are outweighed by burdens of those procedures. When death is inevitable and cannot be prevented by available treatment it is morally permissible to withhold treatment. But limited treatment plan does not equate to no treatment or no care. Medical

professionals are obliged to care for patients, offering palliation as well as emotional support.¹⁸

With this incurable, life threatening and late stage disease, it was no longer relevant to try to treat YN's hypertension and diabetes mellitus to the ideal level to prevent future complications. As long as her blood pressure and blood sugar were not dangerously high, palliation of pain and other symptoms, good nursing care and patient comfort had to be given priority.

Conspiracy of silence

Conspiracy of silence is generally an act of love or a need to protect another from pain. Family members often argue they know the patient better than healthcare professionals. They may argue that telling the truth may take away hopes.¹⁹ However, hiding the truth from YN did not allow her the autonomy to make decisions with regard to specific cancer treatment plan. Withholding the truth prohibits patient and carers from sorting out practical issues, denies them opportunities to reorganize and adapt their lives towards attainment of more achievable goals.²⁰ The best coping strategy would be confrontation of the real situation, open discussions and a good network support for the patient.

4. Palliative care

Delirium

It was thought that YN's sudden change of behaviour was due to hypoglycemia, stress of biopsy procedures or brain metastases. In view of a RBS result of 3.5 mmol/L, a normal head CT scan report, an interval of five days between last biopsy procedure and onset of altered behaviour, it was unlikely these were specific causes. More important factors contributing to her delirium were morphine intake for the first time, several months of uncontrolled pain, worries and anxiety. The stress of going in and out of hospitals with their peculiar strange, unfamiliar faces and surroundings were enough to topple any elderly person into a delirium. Identifying the factors contributing to YN's delirium and rectifying them would be a more useful approach.

Acute hospital care versus home palliative care

Acute hospital care management with forced artificial Ryle's tube feeding, intravenous infusion, in-situ catheterization, were causing tremendous sufferings to YN. These procedures could cause more harm than good. Intravenous infusion can easily lead to fluid overloading and acute pulmonary oedema; catheterization predisposed immunologically incompetent patients to urinary tract infections and septicaemia. Ryle's tube feeding may predispose patient to aspiration pneumonia, may

create a barrier to communication, breathing and talking, not to mention the severe discomfort it incurs in a conscious patient. Confinement to bed weakens the already weak muscles of the elderly and predisposes them to bedsores, pneumonia, sepsis and deep vein thrombosis. Confusion would be even worse! These factors provided explanations as to why YN's delirium did not clear for three weeks in the hospital but she recovered completely once she came home.

The bedrock of palliative care philosophy is that of patient centred holistic care focusing on quality of life and extending support to family and carers. This is encompassed in the following quotation by Dames Cicely Saunders, "You matter because you are you, and you matter until the last moment of your life. We will do all that we can to help you not only to die peacefully, but to live until you die".²¹

A study by J Townsend *et al.* in assessing the preference of terminally ill patients with cancer for their place of final care, showed that majority of them preferred to die in their own home.²² "Most people if given a choice and a supportive home, would choose to die at home and the care of the dying is a traditional part of good general practice. It is a different and rather special form of medical practice and it can be both very demanding and rewarding" (Denis Pereira Gray, President of RCGP).²³ In Malaysia, where palliative care is not well established, preference to die at home may not be an option for many patients even if they wish to do so.

Definition of a good death

YN never recovered from her inability to walk till the end. For the elderly, dependency on others and loss of dignity is more feared than death itself. A study conducted by M Miyashita *et al.* on good-death concept for Japanese cancer care concluded that the main domains on top of the list to define a "good death" are: physical and psychological comfort, dying in a favourite place, good relationship with family and medical staff, maintaining hope and pleasure, not being a burden to others, physical and cognitive control, environmental comfort, being respected as an individual and life completion.²⁴

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