

What Else Can It Be?

In this regular feature we will explore cases of rarer lung conditions and their presentation



Doug and his Breathlessness

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Doug is now 70 years old having retired 5 years ago. He was a self-employed plumber for many years in the local area and was well liked by many in the practice – who had benefited from his skills. Doug lives with his wife who is fit and well at the present time. Doug has some osteoarthritis of his knees and was diagnosed with COPD a year before he retired. Like many people of his age, he had started smoking at the age of 14 years and stopped with a 31-year pack history when he was 45 years old.

Doug had a comprehensive history and examination performed at the time of diagnosis, his chest x-ray and full blood counts were normal and he had good quality spirometry which helped to confirm the diagnosis. Doug had been treated with a long-acting muscarinic antagonist and a long-acting beta agonist/inhaled corticosteroid combination and had attended for annual reviews. His last review suggested no significant change in symptoms or his FEV₁, and he had only had one exacerbation in the past requiring treatment with oral steroids and antibiotics.

Three months later he decided he ought to see the practice as he was feeling gradually more breathless when he was walking upstairs, walking outside with the dog or trying to garden – and was getting some minor chest pains. Examination showed normal regular heart rate and heart sounds; his blood pressure was fine; the FEV₁ had not particularly changed from 3 months previously when reviewed; listening to his chest there were no definite focal findings although the clinician wondered if there was some dullness at the right base and reduced air entry.

The clinician decided to arrange an urgent chest x-ray, ECG and review. The x-ray report suggested a small to moderate right-sided pleural effusion, and Doug was spoken to on the telephone and referred on receipt of the report to the suspected lung cancer clinic with a 2-week wait. He had some blood tests arranged and was booked in to see the clinician 1 week later.

The CT scan with contrast was suggestive of malignant pleural mesothelioma (MPM); after discussion with the multidisciplinary team (MDT) at the hospital a pleural biopsy was arranged.

Commentary

The link between asbestos and MPM was made in 1960, although the first reported case was in 1870.¹ Doug has a well-recognised risk factor (his job as a plumber in the building trade would have involved considerable exposure to asbestos as its use was not banned in the UK until 1999). Asbestos is a naturally occurring fibre used to insulate and fireproof buildings and was commonly used in ceiling and floor tiles, pipe insulation and boilers. There are well-recognised occupations which leave people more at risk (see Box 1). Indeed, for people who were born in the 1940s and worked as a carpenter for more than 10 years, the lifetime risk of mesothelioma is 5.9% and 2% for plumbers, electricians and painters. However, it is not uncommon to detect MPM in people who have no known exposure to asbestos or have partners involved in risky occupations (potentially linked to fibres in clothing).

Mesothelioma takes a long time to develop (often 30–40 years after exposure) and common symptoms are usually non-specific (especially breathlessness, cough and chest pain). The chest pain is usually more localised than cardiac. The latest guidance would also suggest that we should not diagnose without investigations,² and that the NICE guidelines for early diagnosis of malignant lung cancers³ should be adhered to. Hence, we should have a low threshold for arranging a chest x-ray (see Box 2). The chest x-ray can sometimes be normal in this situation so, if clinical suspicion is high, our patients should be referred. (A unilateral effusion as in Doug's case should be taken as malignancy until proven otherwise – renal/liver/cardiac causes are usually bilateral). The clinician might have considered doing screening bloods

Box 1: Those involved in industries using asbestos who are at higher risk of developing mesothelioma

- Carpenters and joiners; painters, decorators
- Plumbers, pipe fitters, heating and ventilation engineers
- Electricians, electrical fitters
- Metal plate workers, shipwrights, riveters
- Labourers in other construction trades
- Sheet metal workers
- Construction and energy plant operatives
- Building inspectors
- Vehicle body builders and repairers
- Metalworking production and maintenance fitters
- Shipbuilding, railway and engineering workers
- People who have worked on DIY projects, particularly Artexing ceilings or working with guttering or insulation materials

to look for other causes of breathlessness (eg, full blood count for anaemia, thyroid function testing for thyroid disease or pro-BNP if cardiac failure is suspected). Similarly, it is widely recognised that a normal ECG does not exclude angina hence, if a suspicion is high, it may be appropriate to consider other tests.

Doug came to see the clinician around a week after being seen by the specialist. He indicated that he had been told that the diagnosis is very likely to be MPM and that he is waiting for a definitive treatment plan and says that he has looked up about MPM on the internet. He is interested to know about options in management.

Commentary

Many patients have access to the internet now (or a friend who can help them when they want more information). Reliable information can usually be found on NHS Choices, patient.co.uk and Mesothelioma UK (<http://www.mesothelioma.uk.com/>) or the British Lung Foundation (<https://www.blf.org.uk/support-for-you/mesothelioma>). There can be medico-legal implications of a diagnosis as some people will be entitled to compensation, and information is available on these patient websites relating to obtaining specialist legal advice. Most clinicians will see very few cases in their professional career (in 2015 there were 2,697 cases in the UK).⁴ For clinicians, a recent BTS guideline has been produced.²

It is worth remembering that Doug may require further investigations (PET or MRI scan). Any biopsies will be reviewed by a pathologist with expertise and, as interpretation is difficult, a second pathologist may be involved. The treatment plan will usually be agreed by a specialist multidis-

Box 2: Symptoms that should refer for urgent chest x-ray (urgent within 2 weeks) if unexplained (one if smoker, two if non-smoker). Note: anyone over 40 years old with unexplained haemoptysis should be referred urgently without a chest x-ray for further investigation³

- Cough
- Fatigue
- Shortness of breath
- Chest pain
- Weight loss
- Appetite loss (new 2015)
- Persistent or recurrent chest infection
- Finger clubbing
- Supraclavicular lymphadenopathy or persistent cervical lymphadenopathy
- Chest signs consistent with lung cancer
- Thrombocytosis (new 2015)

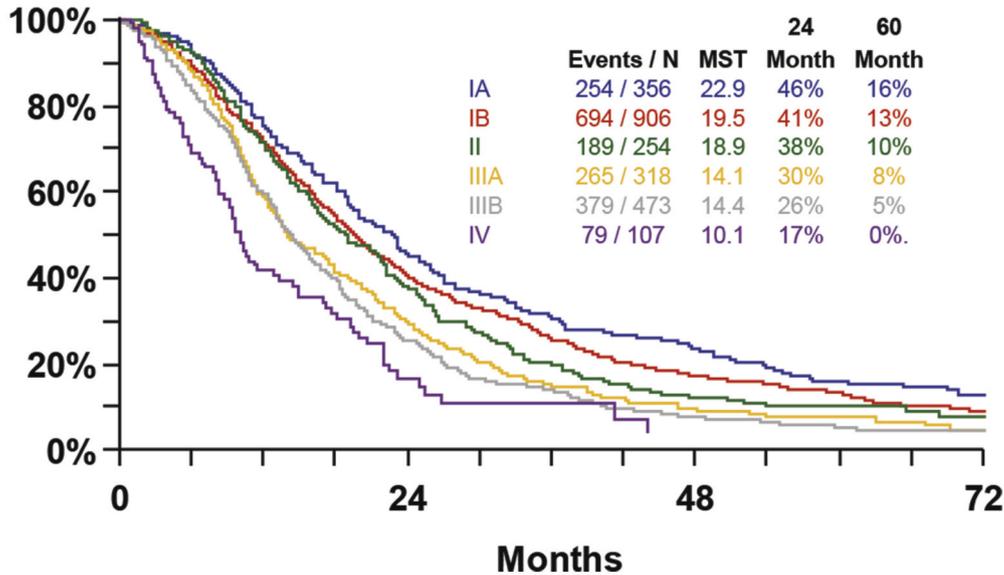
ciplinary team but unfortunately prognosis is not good, with even early disease having a 5-year survival of 16% (Figure 1).⁵

There are several treatments options available:

- Pleural effusions may be managed with talc or indwelling pleural catheters for symptomatic relief (usually not by video-assisted thoracoscopic surgery partial pleurectomy).
- Some people may be appropriately treated with cisplatin and pemetrexed (if good performance status like Doug). The guidance does recommend that, where licensed (not presently in the UK), bevacizumab should be added to this regime.
- Palliative radiotherapy is recommended for localised pain in MPM.
- Symptom control is important and should be managed in keeping with general palliative care – and early involvement is appropriate with specialists if problems with symptoms.

It is important to emphasise to Doug that there will be very specialist care provided in keeping with national guidelines, but that his practice will support him and his wife through this too. The challenge is helping Doug to achieve the best he can in a way that helps him to understand realistically the options available to him, and provide him with the best

Figure 1: Overall survival according to best stage (proposed eighth edition).



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choices possible for his situation. MPM remains a challenge clinically and the prognosis remains poor, but optimising care remains a realistic goal. It is worthwhile planning for the future too with advance directives, ensuring wills are sorted out and considering a lasting power of attorney. Also, as MPM is classified as an industrial disease, it will need to be referred to the coroner who may request a post-mortem and will hold an inquest. The inquest will often delay formal release of a death certificate although may well not delay the funeral plans.

References

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