A Massive Pleural Effusion: What Lies Beneath?

A 21-year-old male of Chinese ethnicity attended the institution’s radiology department for an outpatient chest radiograph for right lower chest reduced air entry. Concurrently, he had an outpatient computed tomography (CT) intravenous pyelography (IVP) for right renal colic and microscopic haematuria for investigation.

What do the radiograph (Fig. 1) and CT (Fig. 2) images show?
A. A pleural effusion
B. Empyema necessitans
C. Pleural mesothelioma
D. Pleural rhabdomyosarcoma
E. Pleural liposarcoma

Findings and Diagnosis
The chest radiograph (Fig. 1) showed a large right-sided pleural effusion, extending superiorly to the periphery of the right upper zone. Partial opacification of the right middle zone adjacent to the pleural effusion was deemed likely related to compressive atelectasis. The heart was mildly shifted to the left. There was also a subtle widening of the interval between the right ninth and tenth ribs posterolaterally (dashed black line) when compared with that on the left side (dashed white line). The left lung was unremarkable.

On CT (Fig. 2), a large right pleural effusion was again demonstrated. However, there was also a large, heterogeneously enhancing, lobulated soft tissue mass (Fig. 2, indicated by *) which was broad-based against, and appears to arise from, the posterolateral parietal pleura of the lower right hemithorax. No internal fat attenuation was noted. There was an extension of the enhancing soft tissue posterolaterally between the right ninth and tenth ribs (Fig. 2, indicated by the arrowheads), with invasion of the intercostal and overlying latissimus dorsi skeletal muscles. No bony destruction of the right ninth and tenth ribs was seen. Other small enhancing pleural nodules were demonstrated in the anterior aspect of the lower right hemithorax.

On further questioning, the patient had non-mechanical right upper back pain for over a year’s duration. The pain was pleuritic in nature and initially mild, but progressively worsened. This was associated with weight loss. He had also been pyrexial for the past month, and recently had a productive cough for 5 days. Clinical examination of the patient revealed an elevated body temperature of 38.1°C, with stony dull percussion over the right lower zone. He
also had an elevated serum C-reactive protein level (175.4 mg/L [<3.0]), but otherwise his blood results were largely unremarkable.

A pleural tap was performed, which was slightly turbid and orange in colour on gross inspection, with no frank pus or haemorrhage. The pleural aspirate had abundant lymphocytes on cytology. The patient was started empirically on rocephin and doxycycline for the presumptive diagnosis of an empyema. A CT study of the thorax was performed—this again showed the heterogeneously enhancing pleural-based mass with large right pleural effusion, which was deemed suspicious for a primary pleural-based neoplasm, or an atypical infection.

The patient subsequently underwent an ultrasound-guided core needle biopsy of the pleural-based mass. Histologic examination of the mass revealed a tumour composed of spindle cells arranged in fascicles, infiltrating into the skeletal muscle. The spindle cells appeared pleomorphic with a high nuclear-cytoplasmic ratio and mitotic figures were present. The tumour cells showed diffuse cytoplasmic staining for WT1, and also stained positive for desmin, CD56 and myogenin, which are muscle-specific proteins. The overall pathology findings were thus in keeping with a spindle cell rhabdomyosarcoma.

To complete the staging procedure, a positron emission tomography-computed tomography (PET-CT) study was performed (Fig. 3). It showed increased metabolic activity of the known primary pleural-based mass, as well as the smaller pleural nodules—the latter suspicious for pleural metastases. There were also metabolically active lymph nodes in the mediastinum and right supraclavicular regions, suspicious for nodal metastases.

Discussion

Rhabdomyosarcoma is the commonest childhood malignancy of mesenchymal origin. Although usually related to skeletal muscle, it can arise from any site, possibly via pluripotent cells differentiating into neurogenic or myogenic elements. Most cases arise in the head and neck region, followed by the genitourinary system and extremities. Thoracic manifestation of primary rhabdomyosarcoma is rare; to date, there have been only 9 reported cases of pleural rhabdomyosarcoma in the literature.1,2

This case is unusual as the patient attended for an unrelated condition, and the diagnosis was made almost serendipitously. The large pleural effusion, although somewhat unusual for a young adult male, could have been readily attributed to a chest infection, given the patient’s history of fever and raised inflammatory markers. However, there was also a very subtle widening of the posterior right ninth intercostal space when compared to the left (Fig. 1).

On CT, the solid components are much more readily appreciable; this was thus not a mere bland pleural effusion. Two main groups of differentials were offered, in the form of a primary malignancy or atypical infection. Another common cause of abnormal pleural soft tissue is pleural metastases (Fig. 4), although this was deemed less likely in this clinical setting given the patient’s relatively young age and absence of risk factors.

Of primary tumours, possible differentials would include pleural mesothelioma (Fig. 5), liposarcoma (Fig. 6) or a rhabdomyosarcoma. For example, mesothelioma may manifest as an effusion with enhancing nodular solid components which can be severe enough to encase the lung in a “rind-like” fashion.3 It can also be locally aggressive, invading the chest wall, mediastinum and diaphragm. Calcified pleural plaques are present in 20% of cases. However, this diagnosis is unlikely for someone in this age group, particularly in the absence of occupational risk factors.

A liposarcoma may occur in this region, but was effectively excluded on CT given the absence of a fatty component in the mass.4

Fig. 3. PET-CT study showing increased metabolic activity in the dominant solid component in the posterior aspect in keeping with a tumour, as well as pleural metastases anteriorly. PET-CT: positron emission tomography-computed tomography

Fig. 4. On this CT study, there are several irregularly-outlined soft tissue nodules arising from the right pleura (indicated by arrowheads). The multiplicity of the lesions, in conjunction with known renal cell carcinoma (not shown), is compatible with pleural metastases. There is also a large right pleural effusion, with midline shift of the mediastinal structures to the left.
Infections resulting in empyema will usually have a pleural effusion and smoothly thickened enhancing pleura (Figs. 7a and 7b). Although uncommon, when a pleural infection extends out of the thorax into or beyond the chest wall, it is known as empyema necessitans. This is usually secondary to atypical infections, of which the 2 most common are tuberculosis and actinomycosis. Actinomycosis produces proteolytic enzymes, which can create fistulae without regard for tissue planes. Soft tissue thickening which mimics nodular pleural thickening (Fig. 8), as well as bony destruction, can be observed.

Ultimately, the patient required tissue sampling to differentiate between a primary malignancy or atypical infection. In our case, this was performed under image guidance using ultrasound (Figs. 9a and 9b). Generally speaking, image-guided intervention may be performed using fluoroscopy, ultrasound, CT or magnetic resonance imaging, depending on which modality the lesion being...
studied is best visualised upon. In this case, either ultrasound or CT would be viable options. However, ultrasound was opted for as it has several advantages over CT such as the absence of ionising radiation (to both the patient and the interventionist), real-time dynamic imaging and lower cost. A formal, preprocedure ultrasound study would have been useful to evaluate the pleural-based lesion of interest, such as in confirming the presence of a solid mass showing vascularity, or identifying cystic spaces representing abscesses within the mass which were potentially drainable, although this was not performed in our case.

Generic, potential complications of imaging-guided percutaneous intervention are haemorrhage, infection and postprocedure pain. More specific potential complications of the ultrasound-guided biopsy performed in our case would include a pneumothorax, haemothorax or inadvertent injury to the subcostal neurovascular bundle.

Conclusion
This young adult male had a chest radiograph which showed a large right pleural effusion. On CT, there were soft tissue components which extended through the intercostal space. Differentials of a soft tissue sarcoma or an atypical infection (empyema necessitans) were entertained. Ultimately, a diagnosis of spindle cell rhabomyosarcoma was made on histology. The learning point here is that a seemingly simple pleural effusion on radiograph may harbour a sinister underlying lesion, and cross-sectional imaging via CT should be performed, in particular, if there were any worrisome clinical features; this patient, for example, had non-mechanical back pain for over a year.

Acknowledgement
The authors would like to acknowledge Drs Angeline Poh, Wong Kang Min and Lee Su Ann for their help in contributing cases towards this article.

REFERENCES

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