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September 2023 Pulmonary Case of the Month: A Bone to Pick

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History of Present Illness

A 56-year-old man presented acute onset of shortness of breath. He denied cough, fever or other symptoms

Past Medical History, Family History and Social History

- Occasional gout
- No relevant family history
- Never smoked

Medications

- Allopurinol
- Multivitamin

Physical Examination

- Other than tachypnea and mild shortness of breath, no significant abnormalities.

Chest X-ray

An AP chest X-ray was performed (Figure 1).

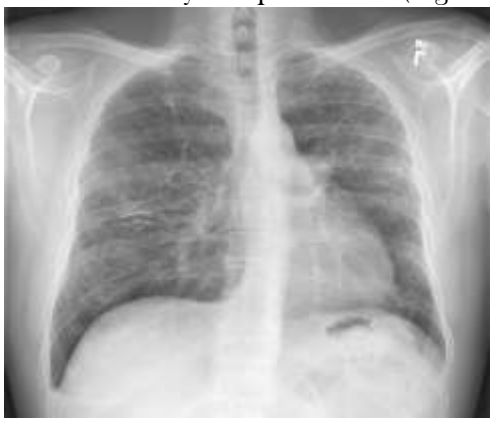

Figure 1. Admission chest X-ray. (Click here to view Figure 1 in a separate, enlarged window)

Which ***abnormality is suggested*** by the chest X-ray?

1. Calcified micronodules in the right lung
2. Retained secretions with atelectasis left lung
3. Right pneumothorax
4. 1 and 3
5. None. The chest X-ray is within normal limits.

Correct!
4. 1 and 3

There is a deep sulcus sign on the right and hyperexpansion of the right hemithorax suggesting a right pneumothorax. A deep sulcus sign may be the only indication of a pneumothorax, especially in a supine film, when air collects anteriorly and basally within the nondependent portions of the pleural space, as opposed to the apex when the patient is upright.

A thoracic CT scan confirmed the presence of the right pneumothorax and the presence of calcified micronodules in both lungs, right greater than left (Figures 2 and 3).

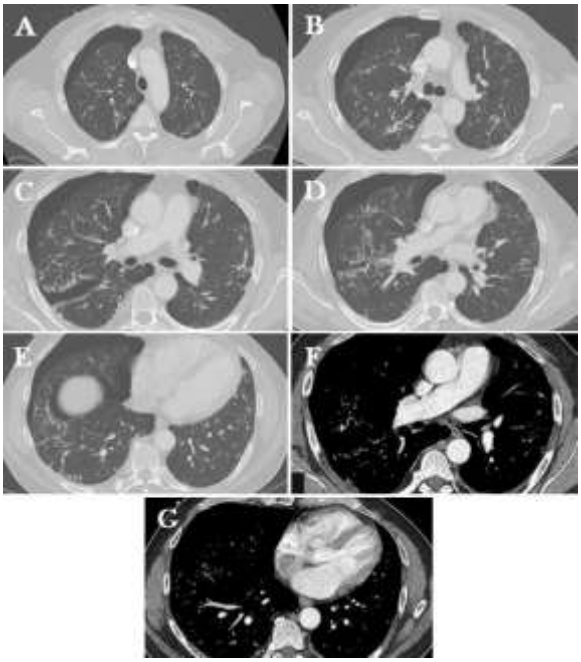

Figure 2. Representative images from thoracic CT scan in lung windows (A-E) and soft tissue windows (F,G). (Click here to view Figure 2 in a separate, enlarged window)

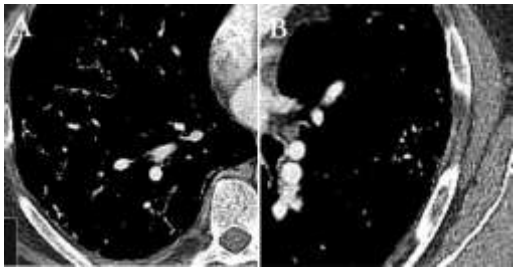

Figure 3. Magnified images from thoracic CT in soft tissue windows from right lung (A) and left lung (B). (Click here to view Figure 3 in a separate, enlarged window)

Which of the following diseases can ***present with a spontaneous pneumothorax***?

1. Connective tissue disease (e.g., Marfan syndrome, Ehlers-Danlos syndrome, rheumatoid arthritis)
2. Asthma
3. Interstitial lung disease (e.g., idiopathic pulmonary fibrosis, sarcoidosis, lymphangioleiomyomatosis)
4. 1 and 3
5. All of the above

Correct!

5. All of the above

Numerous disorders can present with a spontaneous pneumothorax. While primary spontaneous pneumothorax is not associated with underlying pulmonary disease, secondary spontaneous pneumothorax is associated with, but not limited to disorders listed in Table 1 (1).

Table 1. Diseases associated with spontaneous pneumothorax (1).

- Chronic obstructive pulmonary disease
- Asthma
- Cystic fibrosis
- Pneumonia (e.g., necrotizing, Pneumocystis jirovecii)
- Pulmonary abscess
- Tuberculosis
- Malignancy
- Interstitial lung disease (e.g., idiopathic pulmonary fibrosis, sarcoidosis, lymphangioleiomyomatosis)
- Connective tissue disease (e.g., Marfan syndrome, Ehlers-Danlos syndrome, rheumatoid arthritis)
- Pulmonary infarct
- Foreign body aspiration
- Catamenial (i.e., associated with menses secondary to thoracic endometriosis)
- Birt-Hogg-Dube syndrome

(Click here to view Table 1 in a separate, enlarged window)

Which of the following disorders are ***associated with pulmonary ossification***?

1. Sarcoidosis
2. Dendriform pulmonary ossification
3. Pulmonary alveolar microliathsis
4. 1 and 3
5. All of the above

Correct!

5. All of the above

Several disorders can be associated with pulmonary ossification. Pulmonary ossification refers to bone tissue formation (calcification in a collagen matrix), with or without marrow elements, in the lung parenchyma. Table 2 lists several of the disorders to pulmonary ossification (2).

Table 2. Disorders associated with pulmonary ossification.

- Metastatic pulmonary ossification: end stage renal disease, malignancy with hypercalcemia, hyperparathyroidism, hypervitaminosis D.
- Dystrophic calcification: Granulomatous disease, viral infections, amyloidosis, sarcoidosis.
- Pulmonary alveolar microlithiasis: "sandstorm" appearance, due to autosomal recessive gene mutation (SLC34A2).
- Pulmonary ossification-nodular: usually associated with cardiac disorder and venous congestion.
- Dendriform pulmonary ossification: idiopathic or associated with other lung disease.

(Click here to view Table 2 in a separate, enlarged window)

What should be ***done at this time***?

1. Pulmonary function testing
2. Close observation for an expanding pneumothorax
3. Chest tube thoracostomy
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

Pulmonary function testing was performed after resolution of his pneumothorax (Figure 4).

	PREDICTED		CONTROL		POST-DILATOR**	
	Pred	LLN	Actual	%Pred	Actual	%Chng
– LUNG VOLUMES –						
TLC (Pleth) (L)	7.29	5.84	6.16	84		
SVC (L)	5.37	4.48	*3.65	*68		
RV (Pleth) (L)	1.95	1.56	*2.51	*128		
RV/TLC (Pleth) (%)	30.75	24.60	*40.72	*132		
– SPIROMETRY –						
FVC (L)	5.37	4.48	*3.70	*68	*3.78	+2
FEV1 (L)	4.12	3.44	*2.45	*59	*2.72	+11
FEV1/FVC (%)	76.91	64.22	66.39	86	72.12	+8
FEF 25-75% (L/sec)	3.51	1.93	*1.45	*41	2.02	+39
FEF Max (L/sec)	10.18	7.63	*6.59	*64	*7.27	+10
– DIFFUSION –						
DLCOunc (ml/min/mmHg)	30.83	24.66	26.10	84		
DLCOcor (ml/min/mmHg)	30.83	24.66				
VA (L)	7.29	6.09	*5.32	*72		

Figure 4. Pulmonary function testing. (Click here to view Figure 4 in a separate, enlarged window)

A pigtail catheter was placed and the lung reexpanded. In addition, a negative cocci serology and negative ANA, RF and CCP with normal sedimentation rate was observed.

What is the ***diagnosis***?

1. Coccidiomycosis (Valley Fever)
2. Dendriform pulmonary ossification (DPO)
3. Histoplasmosis
4. Pulmonary alveolar microlithiasis
5. Sarcoidosis

Correct!
2. Dendriform pulmonary ossification (DPO)

Based on the clinical presentation, the patient was diagnosed with dendriform pulmonary ossification (DPO). DPO is a result of progressive metaplastic ossification within the lungs (3,4). Typical CT findings include branching nodules which are bone density and usually more prominent in the lower lobes. The branching pattern distinguishes DPO from other causes of lung ossification such as pulmonary alveolar microlithiasis. DPO may occur in association with various lung disorders including IPF, post-ARDS, chronic aspiration and in several pneumoconiosis including asbestosis and heavy metal exposures. Areas of micronodular calcification may also occur in sarcoidosis. DPO can also be idiopathic.

What is the ***treatment of DPO***?

1. Corticosteroids
2. Nintedanib
3. Pirfenidone
4. Roflumilast (Daliresp®)
5. Symptomatic treatment only

Correct!
5. Symptomatic treatment only

In a younger population (<60) with DPO, there is a significant risk of progressive loss of lung function. In older patients it more often is a benign radiologic finding predominantly in men with GERD, OSA or other factors predisposing to microaspiration (3,4). There is no specific treatment other than reducing risk factors (3,4). Our patient was treated with an albuterol inhaler since his PFTs suggested asthma. He has remained stable.

References

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