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Radiologic imaging in Cystic Fibrosis

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ABSTRACT

Cystic fibrosis (CF) is a chronic systemic disease where imaging has long been used for monitoring chest status, and for chest evaluation in exacerbation of the disease. Chest radiography has traditionally been used as the main imaging modality. The abnormal chest radiograph is often perplexing, but abnormal findings can be interpreted easily with a systematic and methodical approach. The companion cases presented this month highlights two basic, but often forgotten or confused findings that have very different meanings: tram-tracks and air-bronchograms.

In the current study authors present two cases along with the assistance of analog imaging phantoms constructed out of household materials, and application of some contrast media. The authors also believe that the presented representative cases of bronchial wall thickening (in the CF case highlighted here) and the finding of tram tracks, compared to consolidation with typical air-bronchograms, should make these findings conceptually clear.

KEYWORDS Cystic Fibrosis, tram track appearance, air-bronchogram.

Introduction:

Cystic fibrosis (CF) is a chronic systemic disease where imaging has long been used for monitoring chest status, and for chest evaluation in exacerbation of the disease. Chest radiography has traditionally been used as the main imaging modality. Computed tomography (CT) is a superior imaging tool compared to radiography [1, 2], but despite its use for more than two decades has yet to show its true clinical value [3, 4]. CT exposes the patient to a substantially higher radiation dose than chest radiography, rendering it unsuitable for the often needed repeated examinations of these patients.

There are countless radiographic "signs" and eponyms involving the common chest x-ray. While many are very specific for a certain disease and others are quite rare, there are a few findings that could be seen on any given day in a busy hospital. The purpose of this article is to provide a clear model for distinguishing two common but key findings, and providing a proper differential diagnosis based on that distinction. Both "tram-tracks" and "air bronchograms" indicate that the airways, normally not visible amongst the lung tissue, can often be traced. The key to this phenomenon is that the pathology for each process is unique, and leads to a separate differential. Historically, the "tram-track sign" has been used as a descriptive radiographic term. For example, there is "tramtracking" of the optic nerve meninges that can indicate a meningioma [5]. Neurology employs a "tram-track" sign that indicates cortical calcifications as seen in patients with SturgeWeber Syndrome [6]. Regardless of the source, "Tram Tracks" are parallel linear densities that stand out from their surrounding tissues. The version discussed below is that of pulmonary tram-tracking, which indicates the visibility of thickened bronchial walls that do not show normal tapering toward the lung periphery. Air-bronchograms, by comparison, do not involve any thickening or inflammation of the bronchioles. Historically, the term "bronchogram" was used to describe a radiograph taken after the inhalation of a radio-opaque substance such as barium sulfate [7]. This would provide an impressive picture of the bronchiole architecture and was used for exploring pulmonary processes prior to the advent of CT imaging. Currently, the commonly used term "air bronchogram" has implication that the "contrast material" is the normal air within the bronchioles. In contrast to the original bronchogram, the bronchioles are only visible when a pathologic process involving the surrounding tissue creates a significant change from the normal lung density and highlights the lucency of the air-filled bronchioles.

Aim and Objectives:

Current study aimed to illustrate the differences between tram-tracks and air-bronchograms, in an experimental model.

Material and Methods:

To illustrate the differences between tram-tracks and air-bronchograms, an experimental model was created. Household items including kitchen sponges soaked in both tap water and IV contrast material, an apple, and straws of various diameter were imaged using both plain film x-rays as well as a CT scanner. The water-soaked sponges provide the baseline "lung tissue," allowing visualizing a portion of the architecture, but with most of the image being radiolucent. Straws placed into the sponges had varying appearances depending on the diameter of the straw and the thickness of the straw wall. To create "bronchiole thickening" it was necessary to wrap the straws with several layers of medical tape soaked with contrast. An apple was used in place of the sponge to simulate lung tissue that was consolidated. Straws pushed through the apple appeared as tubular lucencies when contrasted to the dense surrounding tissue, and there was no appreciable visualization of the straw itself. These models will provide the analog images used in this article.

Observations:

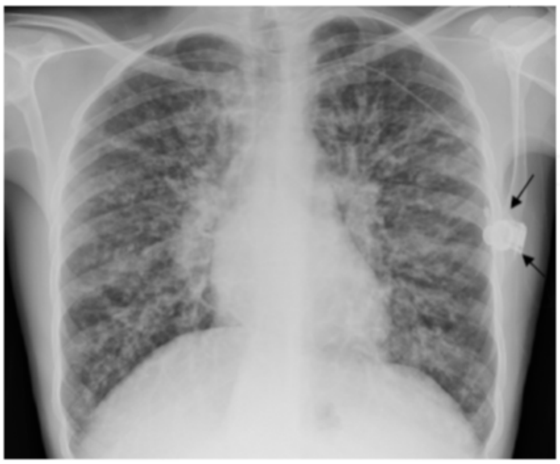

Fig 1: Chest radiograph of a CF patient with hyperinflated lungs and severe bilateral and diffuse bronchiectasis seen as ring shadows and tram-track opacities that converge to the lung hila, which correspond to dilated thick walled bronchi.

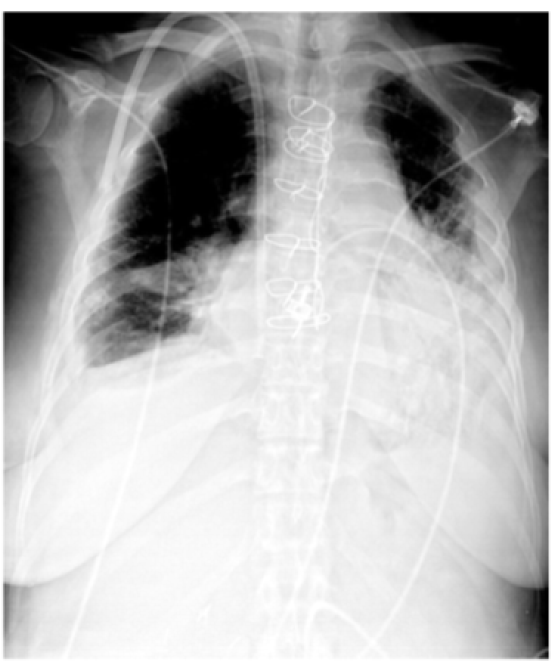

Fig 2: PA Chest X-Ray of a 53 year old post-operative woman who displays Air Bronchograms bilaterally. The left lower lung field contains a "complete air bronchogram," meaning the tubular lucency extends from hilum to periphery. The right lower lung shows a less obvious air bronchogram exiting the hilum inferiorly.

The first case highlighted as the unknown image involves a patient with cystic fibrosis, a well-known entity that involves mucus plugging of the airways, chronic bronchiole inflammation and infection, and ultimately diffuse bronchiectasis. Figure 1 shows multiple radiographic findings consistent with the pathophysiology of cystic fibrosis. Mucus plugging of the small bronchioles results in hyperinflation of the lungs. There are multiple nodular opacities which represent the mucus plugs. The aforementioned tram-tracking is visible in all lung fields, most conspicuous in the bilateral upper lung fields and extending laterally from the right hilum. In addition, multiple ring-shaped shadows can be seen scattered throughout. This patient is a long-time sufferer of cystic fibrosis, and represents the example of a radiograph with tramtracking secondary to bronchiectasis.

The second case for comparison purposes is presented and is an old woman 2 days status-post thoracotomy who developed shortness of breath. This will be used as the air-bronchogram example. As the differential diagnosis for air-bronchograms is extensive, only superficial coverage of the disease entities mentioned will be provided. Figure 2 shows diffuse, bilateral lower lung field densities. The left heart border is obscured, as are both costophrenic angles and hemi-diaphragms. The sternal wires, ECG leads, and surgical drain are visible, and are consistent with the patient's history as a post-operative patient. Of the greatest importance for this discussion is the presence of the complete air-bronchogram in the left lower lung field and the partial air-bronchogram in the right lower lung field. The complete air-bronchogram can be seen to extend from the hilum out into the peripheral lung field, and outlines several branches of the bronchial tree. This case most likely represents atelectasis due to prolonged immobilization.

Discussion:

A thorough understanding of the mechanisms involved in creating either the tram-track sign or air-bronchogram leads naturally to creating a reasonable general differential diagnosis. Often times, the clinical features combined with the radiographic findings can lead to a fairly specific diagnosis.

Figure 3: Tram-tracking in the experimental model, with plain film to the left, and axial CT on the right. The plain film shows one straw in profile (left) and one straw in oblique (right). The CT image shows the increased attenuation of the tape-wrapped straw mimicking bronchial wall thickening.

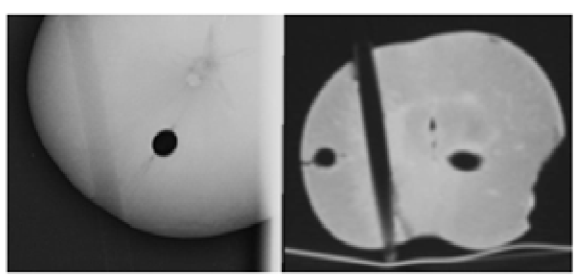

Figure 4: Experimental model of Air Bronchogram using the Apple/Straw setup. On the left is the plain film version showing one straw in profile (the air bronchogram), and one on end illustrating the potential for image variation depending on angle. On the right, the CT reconstruction, with the addition of a second straw in oblique (oval near midline) orientation.

Tram-tracking was presented in the experimental model as a thick-walled tube in a normal surrounding. How does this situation evolve in a patient? To create a biologic tram-track, two processes need to occur. The first is bronchiole dilation, and the second is bronchiole inflammation. Bronchiole dilation occurs readily when obstruction of the airways leads to air-trapping and mechanical stretching of the airway. Likewise, weakening of the muscular layers of the bronchioles leads to an unbalancing of the forces that maintain the airway. The outward pull of the elastic lung parenchyma causes unopposed bronchiole dilation [8]. The inflammatory component can be either acute or chronic. Acute inflammation, as in any other area of the body, leads to edema of the bronchiole. Chronic or recurrent inflammation can have a more lasting effect, causing permanent damage to the muscular and elastic layers of the airway. Additionally, damage to the ciliary layer predisposes to further obstruction and infection, setting the stage for a worsening cycle of lung injury.

As this article has shown, two of the more common chest X ray findings, tram-tracking and air bronchograms, can be confusing to identify and distinguish. With a thorough understanding of their pathophysiology and differential diagnoses, a variety of conditions and diseases can be either excluded or included based on a particular radiograph's appearance.

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