Abstract

Spontaneous pneumomediastinum (SPM) is described as free air or gas located within the mediastinum that is not associated with any noticeable cause such as chest trauma. SPM has been associated with many conditions and triggers, including bronchial asthma, diabetic ketoacidosis, forceful straining during exercise, inhalation of drugs, as well as other activities associated with the Valsalva maneuver. The Macklin effect appears on thoracic computed tomography (CT) as linear collections of air contiguous to the bronchovascular sheaths. With the recent availability of multidetector-row CT, the Macklin effect has been seen in the clinical setting more frequently than expected. The aim of this review article is to describe the CT imaging spectrum of the Macklin effect in patients with SPM, focusing on the common appearance of the Macklin effect, pneumorrhachis, and persistent SPM with pneumatocele.

INTRODUCTION

Pneumomediastinum is described as free air or gas located within the mediastinum. It can be precipitated by various triggers that are either intrathoracic, such as stenosis or blockage of an airway, Valsalva maneuver, blunt trauma to the chest, or ruptured alveoli; or extrathoracic, such as fractured sinus, iatrogenic manipulation during tooth extraction, or ruptured intestine.[1]

Spontaneous pneumomediastinum (SPM) is described as free air or gas located within the mediastinum that is not associated with any noticeable cause such as chest trauma. The first case series of SPM was reported by Hamman[2] in 1939; therefore, the condition is called Hamman syndrome.[3] Respiratory pneumomediastinum is a result of rupture along the alveolar tree, which leads to an abrupt increase in the intra-alveolar pressure. Released alveolar air centripetally dissects through the pulmonary interstitium along the bronchovascular sheaths.
toward the pulmonary hilum, into the mediastinum, and subcutaneous emphysema due to ruptured pars membranosa (arrow). This pathophysiological mechanism was described by Macklin and Macklin in 1944, and is known as the Macklin effect.

SPM is usually a benign, self-limiting illness affecting young males. However, it is a condition that is not widely recognized by clinicians. There have been several reports describing the appearance of the Macklin effect on computed tomography (CT) images of patients with SPM[5-12]. This review article will describe the CT imaging spectrum of the Macklin effect as observed in patients with SPM.

The Main Causes of SPM

SPM occurs predominantly in young males[13,14], and is an uncommon entity. The prevalence of SPM reportedly ranges from 1 in 8005 to 1 in 42,000 hospital admissions and emergency admissions[13,15]. Three different mechanisms can produce pneumomediastinum: (1) disruption of a cutaneous or mucosal barrier (usually the tracheobronchial tree or the esophagus), which allows the entry of gas into the mediastinum; (2) gas produced by organisms in the mediastinum or adjacent chest; or (3) rupture of an alveolus. Alveolar rupture is known as SPM of an alveolus. Alveolar rupture is known as SPM of an alveolus. This article will describe the CT imaging spectrum of the Macklin effect as observed in patients with SPM.

CT Demonstration of the Macklin Effects in SPM

Macklin and Macklin first observed that released alveolar air from alveolar rupture centripetally dissected through the pulmonary interstitium along the bronchovascular sheaths toward the pulmonary hilum and into the mediastinum[16]. Wintemark and Schnyder recently reported that the rate of Macklin effect seen on chest CTs of patients with blunt trauma to the chest was 39%. They concluded that CT-associated Macklin effect was a sign of severe blunt trauma to the chest[18]. However, there have been several reports of the Macklin effect on the CT scans of patients with SPM[5-12].

As demonstrated in Figures 2-5, the Macklin effect appears on thoracic computed tomography (CT) as linear collections of air contiguous to the bronchovascular sheaths[5-12]. The air dissected into the pulmonary hilum and from there enters the mediastinum. We previously reported that using multidetector-row (MD)CT, we detected the Macklin effect in 8 of 9 patients with nontraumatic pneumomediastinum, which was a higher rate of detection than had been previously reported[19]. Sakai et al[20] also reported a high detection rate of the Macklin effect using 64-detector-row CT. They found interstitial gas in the perihilar region of all 20 of their patients. We speculated that the increased detection rate of the Macklin effect was a result of using MDCT with application of thin collimation, a one-breath-hold technique, and visualization of magnified images on a monitor with cathode ray tubes. These factors might facilitate the identification of subtle Macklin effects. Therefore, we may conclude that
alveolar rupture described as the Macklin effect is even frequently seen in patients with SPM.

CXR are generally useful for diagnosing pneumomediastinum, although there have been false-negative results. For false-negative cases, Okada et al concluded that because of thin slices obtained on CT, CT is more effective than CXR alone for diagnosing pneumomediastinum. Sixty-four-detector-row CT reveals minute changes in organs and peripheral tissues. However, the Macklin effect was not detected in the peripheral lung of 4 of our reported 12 cases and in 11 of 20 cases in Sakai’s report. We believe that since the Macklin effect develops as linear collections of air in the pulmonary interstitium that extend along the bronchi and contiguous blood vessels to gradually reach the perihilar bronchovascular sheath, the longer that time passes after its onset, the less often it is seen in the periphery of CT scans (Figure 3).

Complications of SPM

SPM is occasionally associated with pneumorrhachis, the presence of air within the spinal epidural space (Figure 4). A literature review of 48 patients with pneumorrhachis revealed that only 1 case had neurologic symptoms and signs; the other cases were successfully managed conservatively. This literature review described a 72-year-old man with progressive motor weakness and sensory deficits in the lower extremities, who had a large accumulation of intraspinal air. He recovered completely after a C7 laminectomy. Kono et al reported pneumorrhachis in 4 of 42 children with SPM, and the patients with pneumorrhachis did not have neurological symptoms. Therefore, in SPM, a collection of air within the spinal canal is mostly self-limiting and benign. Pneumomediastinum concomitant with pneumoperitoneum is very rare in SPM, with only a few cases reported. It also appears to resolve with conservative treatment, without intervention.
Although the Macklin effect appears on thoracic CT as linear collections of air contiguous to the bronchovascular sheaths, the onset, which is alveolar rupture, is rarely observed on CT. The released alveolar air rapidly dissect into the pulmonary hila and from there enters the mediastinum. We did have an SPM patient with a pneumatocele (Figure 5). This young female patient had interstitial pneumonia with prolonged SPM and cervical subcutaneous air. Patients found to have a Macklin effect involving peribronchovascular air and pneumatocele[30] will have a prolonged SPM, and clinical intervention is required.

CONCLUSION

The Macklin effect can be frequently observed on the MDCT images of patients with SPM not associated with trauma. A Macklin effect seen on CT may help differentiate respiratory from other etiologies of pneumomediastinum. However, especially in pediatric patients with SPM who improve clinically, aggressive investigation and follow-up CXRs are rarely warranted, and the efficacy of CT is limited.

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