Pneumomediastinum, pneumorrhachis, and subcutaneous emphysema in *Pneumocystis jiroveci* pneumonia in AIDS

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Pneumomediastinum, the presence of free air within the mediastinal cavity, is sometimes accompanied by subcutaneous emphysema and pneumorrhachis (air within the spinal canal). We report the case of a 28-year-old man with previously undiagnosed HIV who was diagnosed with extensive pneumomediastinum, pneumorrhachis, and subcutaneous emphysema secondary to *Pneumocystis jiroveci* pneumonia after presenting with chest pain, dyspnea, and central cyanosis. Surgical consultation was requested, but a conservative approach of observation proved sufficient as the free air was resorbed into the surrounding tissues.

P*neumocystis jiroveci* pneumonia is a common opportunistic infection in patients with AIDS and low CD4 counts <200 cells/mm³. In most cases, *P. jiroveci* pneumonia presents with chest pain, cough, shortness of breath, and hypoxemia. It has a high mortality rate unless treated promptly with antibiotics and, if severe, steroids. Pneumatocoles that form as a complication of *P. jiroveci* pneumonia can rupture, allowing air to leak and spread along the peribronchial linings and facial planes of the chest, neck, and axilla to give rise to subcutaneous emphysema, pneumomediastinum, and in some severe cases pneumorrhachis (1–3). Noninvasive imaging studies such as chest x-ray and computed tomography (CT) scan of the chest are usually enough to detect these findings. Pneumomediastinum, pneumorrhachis, and subcutaneous emphysema usually resolve spontaneously, and a conservative strategy of close monitoring and observation may be sufficient.

CASE REPORT

A 28-year-old man with no significant past medical history presented to the emergency department with substernal chest pain, dyspnea, orthopnea, and central cyanosis of 1 week duration. The symptoms progressively worsened and became very severe the night prior to his presentation. He reported fever, malaise, fatigue, and mild cough productive of clear, whitish sputum. He denied any trauma, allergies, sick contacts, recent travels, or similar episodes in the past. He reported a weight loss of about 90 lbs over the preceding year. The patient reported that he had not been sexually active for the previous 9 months but had been sexually active with both men and women and used barrier protection inconsistently. On examination, he ap-

peared in moderate to severe distress and had central cyanosis, with a pulse of 109 beats/minute, blood pressure of 132/79 mm Hg, temperature of 97.5°F, respiratory rate of 26 breaths per minute, and oxygen saturation of 76% on room air. He was placed on a nonrebreather mask, and his oxygen saturation improved to 96% to 99%. There was minimal crepitus on palpation of the neck and chest wall and scattered crackles throughout both lungs.

Chest radiographs revealed diffuse bilateral opacities, marked subcutaneous emphysema in the neck, and pneumomediastinum (*Figure 1a*). A subsequent CT scan of the chest confirmed these findings and also showed emphysema in the axilla and pneumorrhachis (epidural pneumatosis) in the thoracic spinal canal (*Figures 2*). The patient's HIV test was positive. Given the high suspicion for pneumocystis pneumonia in the presence of HIV infection, the patient was started empirically on intravenous trimethoprim-sulphamethoxazole and prednisone. Over the following 2 days, the dyspnea and chest pain disappeared. He was transitioned to nasal cannula oxygen. Subsequent chest radiographs revealed significant improvement in the extent of subcutaneous emphysema and pneumomediastinum (*Figure 1b*).

Once stable, the patient underwent bronchoscopy, and bronchial washings confirmed the presence of *P. jiroveci* pneumonia on Gomori methenamine silver staining. Direct fluorescent antibody on the bronchial washings also confirmed the presence of *P. jiroveci* pneumonia. The patient had an HIV viral load of 485,425/mm³ and CD4 count of 24. On day 5 of his hospitalization, he was switched to oral trimethoprim-sulphamethoxazole and prednisone and was discharged in stable condition to continue outpatient follow-up for reassessment and preparation for initiating highly active antiretroviral therapy.

DISCUSSION

Spontaneous pneumomediastinum—the presence of free air in the mediastinum without an obvious etiology—occurs in 1 in 30,000 emergency department visits (1, 2). The co-occurrence

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Figure 1. (a) Chest x-ray showing bilateral pulmonary infiltrates with prominent symmetrical pneumomediastinum (black arrow) associated with subcutaneous emphysema in the base of the neck and along both shoulders (white arrows) and axilla (white arrowhead). (b) Chest x-ray, day 3, showing decreased amount of pneumomediastinum (white arrow) and subcutaneous emphysema in the soft tissues of the neck. Air in the axillary regions is almost totally resorbed (white arrowhead).

of spontaneous pneumomediastinum and pneumorrhachis is rare, and few cases have been reported (3–5).

Spontaneous pneumomediastinum occurs with abrupt transalveolar pressure gradient increases like a Valsalva maneuver, violent coughing, or intense physical exertion. The pathophysiologic process was described by Macklin in 1939 (6). The Macklin effect refers to an alveolar rupture with air dissection along the interstitial sheaths toward the mediastinum, eventually leading to a pneumomediastinum (7). Alveolar rupture is responsible for more than 95% of cases and is a more frequent cause than esophageal and tracheobronchial disruptions (7).

The clinical presentation of pneumomediastinum is benign and self-limiting. The most common complaints are chest pain and dyspnea; chest pain is commonly pleuritic and retrosternal. CT of the chest is the gold standard for diagnosis. If suspicion for aerodigestive tract perforation is high, a barium swallow,

esophagoscopy, or bronchoscopy may be performed. The management of patients with spontaneous pneumomediastinum is conservative, consisting of rest, analgesics, and close observation. The utility of antibiotics, oxygen therapy, and restriction of oral intake is debatable (1).

Although pneumomediastinum is benign, the risk of tension pneumomediastinum or pneumothorax justifies close clinical observation. Pneumopericardium may also occur, with complications such as air tamponade and cardiac herniation (8, 9). For the management of pneumomediastinum, Okada et al (8) proposed that patients without (i) fever $<38^{\circ}\text{C}$, (ii) oxygen desaturation $<96\%$, (iii) progressive symptoms, (iv) vomiting at the onset, and (v) anxiety should receive ambulatory care.

Pneumorrhachis is usually an asymptomatic radiologic finding (9). Pneumorrhachis can be classified into traumatic, nontraumatic, and iatrogenic types (4). Pneumorrhachis usually

Figure 2. (a) Chest CT scan and (b) mediastinal window showing bilateral ground-glass infiltrates, subcutaneous emphysema in the lateral chest wall and axillary region (white arrowhead), free air surrounding mediastinal vascular structures (blue arrow), massive pneumomediastinum (white arrow), and free air in the thoracic spinal canal (black arrow).

does not cause any symptoms and is commonly detected incidentally. Pneumorrhachis without associated trauma or intervention is rare (9, 10). There are few reports of combined pneumomediastinum and extradural pneumorrhachis not associated with trauma (9, 10). Our patient developed pneumorrhachis, pneumomediastinum, pneumothorax, and surgical emphysema in the absence of trauma. In trauma victims, these findings may suggest spinal fracture or hidden severe injuries. Pneumorrhachis can be diagnosed on a radiograph or a CT scan of the spine, with CT being preferred (4, 11).

The accumulated air may cause symptoms like mild radicular pain, neurological compression, and sequelae such as paraplegia (9, 10, 12). Spontaneous pneumorrhachis does not require a specific intervention, as most cases are self-limiting. Spontaneous resorption of the air within 2 to 3 weeks commonly occurs (8–10). Surgical intervention can be considered when air acts as a space-occupying lesion, causing pressure on nervous structures and resulting in neurological deficits (8–10).

Pneumomediastinum is a rare complication of *P. jirovecii* pneumonia and has been reported at the time of diagnosis, mechanical ventilation, and trimethoprim-sulfamethoxazole treatment (11, 13, 14). In our case, we gave antimicrobial therapy for *P. jirovecii* pneumonia early and adopted a conservative strategy. Our case is unique as the patient, with previously undiagnosed HIV/AIDS, presented with extensive pneumomediastinum, pneumorrhachis, and subcutaneous emphysema caused by *P. jirovecii* pneumonia. Although pneumomediastinum has been described in relation with *P. jirovecii* pneumonia, concurrent pneumorrhachis related to the same etiology has rarely been reported.

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