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Case Report

Cerebral air embolism associated with cavitary lung lesion in a patient with Ehlers-Danlos syndrome ☆

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ABSTRACT

Pneumocephalus, is a general term describing the presence of air within the intracranial structures. It most commonly occurs due to dural injury, often the sequela of head trauma or surgery. More infrequently, nontraumatic pneumocephalus can be related to infection, Valsalva, fistulization between air-containing organs and intracranial structures, or vascular air embolism. While postsurgical pneumocephalus is often benign, serious consequences of pneumocephalus exist, including tension pneumocephalus and cerebral infarction. We present a case of air embolism and cerebral infarction in a patient with Ehlers-Danlos syndrome, found to have large cavitary lesion in the left upper lobe of the lung, with associated pulmonary vascular malformation seen on bronchoscopy. To our knowledge this is the first reported case of air embolism associated with a pulmonary cavitary lesion and vascular malformation, in a patient with Ehlers-Danlos syndrome.

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Introduction

Intracranial air, also known as pneumocephalus [2], generally occurs as a result of injury to the dura, often following head trauma, diagnostic procedures, or surgery [4], and less frequently in the setting of Valsalva and changes in atmospheric pressure. Air embolism is a rare cause of pneumocephalus, with high morbidity, often complicated by cerebral infarction [3]. As with other etiologies of pneumocephalus, air embolism usually occurs following trauma, surgery, or diagnostic procedures; however, it can result from any cause of air entry into the systemic vasculature. We present the first reported case of air embolism and cerebral infarction in a patient with Ehlers-Danlos syndrome, found to have large cavitary lesion in the left upper lobe of the lung.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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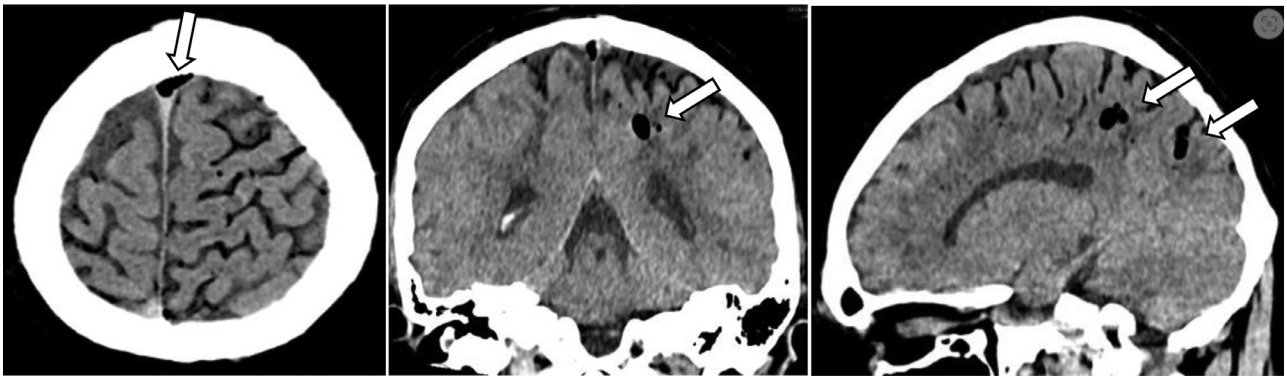

Fig. 1 – Noncontrast enhanced CT axial, coronal, and sagittal images of the head demonstrating air in the superior sagittal sinus and in the left frontoparietal subarachnoid spaces, as demonstrated by the white arrows.

Case

Patient is a 22-year-old male with autism and Ehlers Danlos syndrome, who was brought to the emergency room by his mother after she found him rigid, shaking, and unresponsive. Patient had been watching television in his room prior to when his mother found him. Upon arrival at the ER, patient had a National Institute of Health Stroke Scale (NIHSS) score of 28, with pinpoint pupils and intact brainstem reflexes. Patient did not respond to Narcan administration. A stroke code was called, and noncontrast enhanced CT of the head demonstrated extensive bilateral air emboli (Figure 1). CT angiography (CTA) of the head and neck with intravenous (IV) contrast showed a cavitary lesion in the left lung apex containing an air-blood level, and surrounding ground glass opacities, measuring approximately 2.8 × 2.3 cm (Figure 3). Subsequent noncontrast enhanced brain MRI showed diffuse bilateral infarction (Figure 2). Once stabilized, bronchoscopy was performed which identified a pulmonary vascular malformation at the site of lung cavitation, thought to be a possible source of the air embolism. CTA of the chest with IV contrast did not identify a corresponding pulmonary arteriovenous malformation, but did demonstrate significant ectasia of the main pulmonary artery, measuring 5.0 cm in diameter (Figure 4). Echocardiogram was performed which did not find evidence of patent foramen ovale, and extensive infectious disease work-up was negative.

Patient's hospital course was complicated by sustained increased intracranial pressure, requiring ventriculostomy, and prolonged intubation requiring tracheostomy and percutaneous gastrostomy tube placement. Patient was eventually discharged to long term acute care hospital for ventilator weaning.

Discussion

Pneumocephalus is a general term describing the presence of air within the intracranial spaces [2]. It occurs primarily following injury to the dura. Common causes include head trauma, surgery, and certain diagnostic procedures [4]. Less commonly, nontraumatic pneumocephalus can occur due to infection by gas-forming organisms, and increases in intracranial pressure secondary to variations in atmospheric pressure and Valsalva. In more rare cases, pneumocephalus is related to fistulization between air-containing organs and intracranial structures [2] or from vascular air embolism.

In certain cases, such as after surgery, pneumocephalus may be asymptomatic; however, serious complications can occur, including tension pneumocephalus [5], and in the case of air embolism, cerebral infarction.

Our case is of particular interest as it represents an example of air embolism associated with a pulmonary cavitary lesion and vascular malformation in a patient with Ehlers-Danlos syndrome. While cerebral infarction due to air embolism has been reported in a patient with a lung cavitary lesion in the setting of pulmonary tuberculosis [10], in our case, an extensive work-up did not find any evidence of tuberculosis or other infectious cause for pulmonary cavitation. To our knowledge, this is the first reported case of air embolization in the context of cavitary lesion in Ehlers-Danlos syndrome.

Ehlers-Danlos is a spectrum of heritable connective tissue syndromes, related to each other by presence of defects in the genes involved in structure and synthesis of collagen proteins [6]. Hallmark symptoms include tissue fragility, joint hypermobility, and skin hyperextensibility. The manifestations of Ehlers-Danlos syndrome can be quite variable given the ubiquity of collagen in the human body [6].

Vascular Ehlers-Danlos Syndrome (vEDS) is an umbrella term which describes subtypes of EDS typified by vascular fragility, whose patients are predisposed to vascular events, including dissection and aneurysm formation [7]. Presence of significant pulmonary arterial ectasia in our patient is most consistent with vEDS.

Different pulmonary pathologies and lesions have also been reported in patients with vEDS, including formation of cavitary nodules [8]. These are thought to develop as a sequela of spontaneous pulmonary hemorrhage and hematoma formation [9].

In our case, the overlap of the pulmonary and vascular syndromic manifestations of vEDS are likely to have led to the formation of the cavitary lesion in the patient's left lung apex,

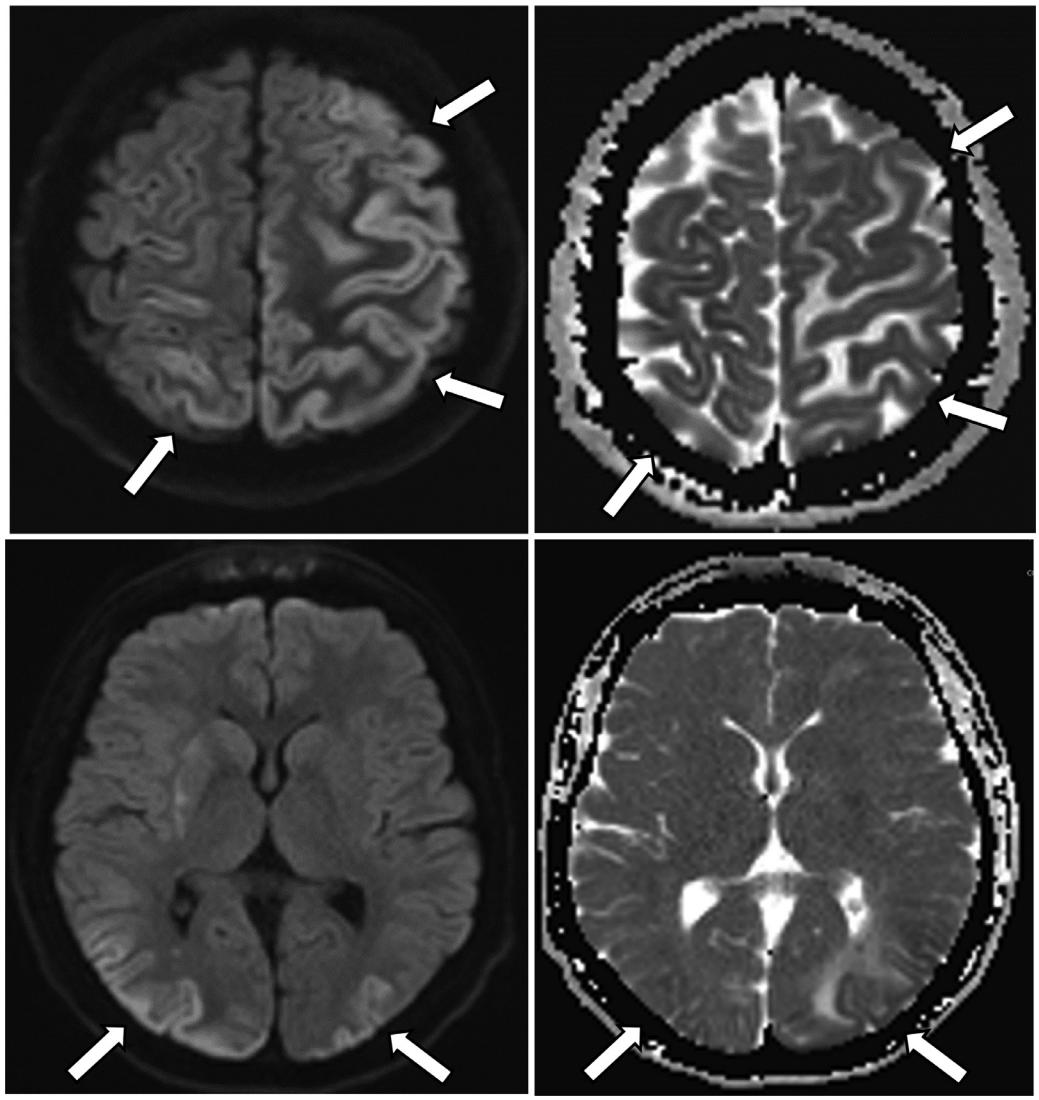

Fig. 2 – Axial MR, DWI, and ADC images demonstrating diffuse bilateral acute/subacute infarcts, as demonstrated by the white arrows. Pattern of cortical restricted diffusion with subcortical white matter vasogenic edema has been described in another case of cerebral air embolism [1].

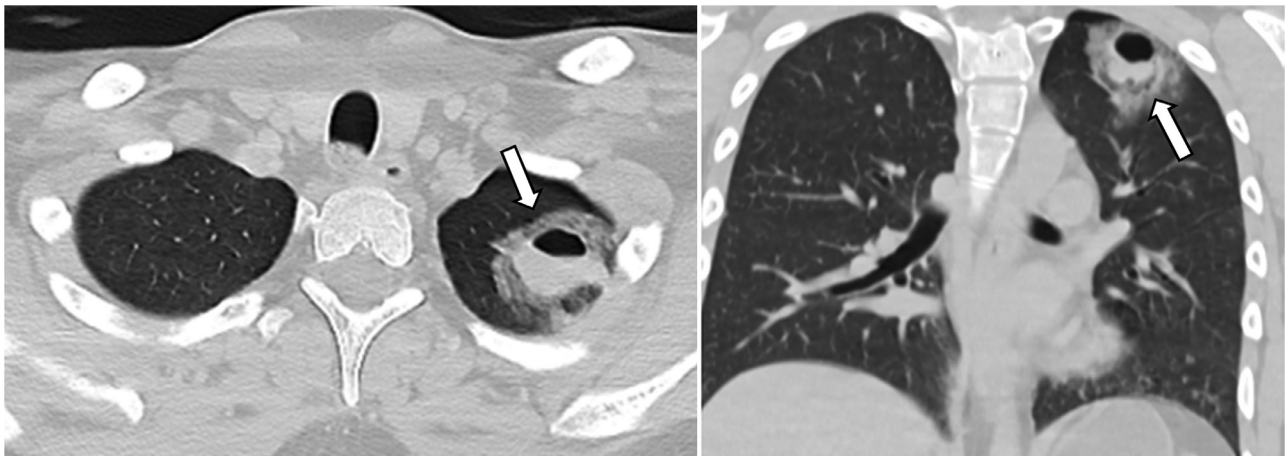

Fig. 3 – CTA of the chest with IV contrast in lung window demonstrates left apical cavitary lung lesion containing hemorrhage, and blood-fluid level, as demonstrated by the white arrows. There are surrounding perilesional ground glass opacities.

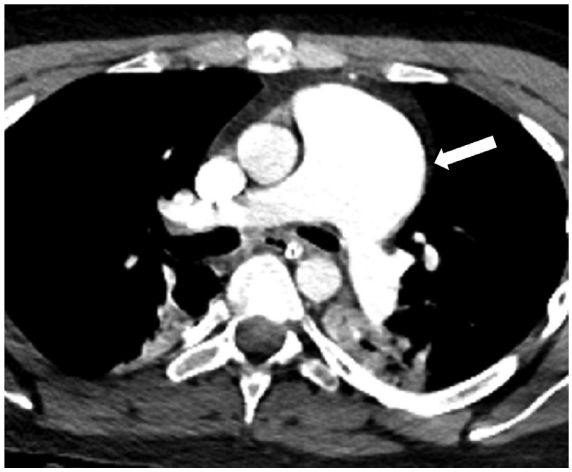

Fig. 4 – CTA of the chest with IV contrast demonstrates significant ectasia of the main pulmonary artery, measured to approximately 5.0 cm in diameter, as demonstrated by the white arrow.

resulting in pulmonary vascular injury and the development of the vascular malformation seen on bronchoscopy. While corresponding arteriovenous malformation was not identified on CTA of the chest with IV contrast, a number of possibilities may explain this, including: 1) presence of broncho-venous fistula, 2) arteriovenous component in the vascular malformation below the resolution of CTA, 3) transient broncho-arterial fistulization within the vascular malformation, 4) migration of air into the pulmonary capillary bed facilitated by damage to the wall of the cavitary lesion [10], without component of arteriovenous or bronchoarterial fistulization.

Although beyond the scope of this case report, it would be interesting to further determine if pulmonary cavitation associated with EDS is a significant risk factor for air embolism, and if stratification of risk based on EDS status can improve patient outcomes.

Conclusion

Cerebral infarction is a devastating complication of air embolism. We present a case of air embolization associated with lung cavitation in the setting of Ehlers-Danlos syndrome. To our knowledge, this is the first reported such case.

Patient consent

Consent for publication of the case was obtained from the patient.

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