

with transthoracic needle biopsy or marking of the lung is estimated at 0.002% [3]. This case seems to be the first report of a cerebral air embolism developing after anti-cancer agent administration through a chest tube.

The first clinical characteristic of a cerebral air embolism is that it occurs at the right side of the cerebral area because air can easily flow into the first branch of the aortic arch, which is the right brachiocephalic artery. Afterward, the bubble is usually observed to disappear within 24 hours on brain CT scan or magnetic resonance imaging. In almost all cases in the literature, the subject recovered neurogenic disturbance. However, in our case, a left-sided hemiplegia continued until death, although consciousness returned within 24 hours. Changes in the findings on CT scan and magnetic resonance imaging well reflected the clinical sequence from the detection of the bubble to the development of the ischemia, edema, and infarction. When such a cerebral air embolism occurs, the patient's head should be lowered as quickly as possible under oxygen supply. A hyperbaric oxygen unit should be transferred if possible [4].

Two mechanisms are hypothesized to explain why a large amount of air existed in the peripheral pulmonary vein and then in the systemic circulation. First there was a possibility that the tip of the chest tube that directly contacted the visceral pleura had advanced further into the pulmonary tissue. Of course it was ensured that the tip of the tube was outside the lung when drainage was done. Next the inflammation may be induced by the anti-cancer agent to the visceral pleura [5]. The inflammation may have formed a fistula between the peripheral bronchus and the pulmonary vein. The increased pressure gradient caused a great deal of air to be inhaled into the pulmonary vein through the fistula by means of coughing, and the air then entered the systemic circulation. The second mechanism seems most likely in our case, because air was never expelled from the chest tube during the entire drainage period. Care must be taken to be sure that the tip of the chest should not vertically contact the visceral pulmonary pleura when drugs are scheduled to be administered. A dose of drug should be determined with caution, because the concentration of drugs would markedly increase in a limited space such as a pleural cavity with adhesion.

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Diagnosis of Birt-Hogg-Dube Syndrome in a Patient With Spontaneous Pneumothorax

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Birt-Hogg-Dube syndrome refers to a dermatologic syndrome, consisting of small papular skins lesion distributed on the scalp, forehead, face and neck, which is autosomal dominantly inherited. Subsequently patients may develop concomitant renal and thoracic pathology. We report the case of a patient with Birt-Hogg-Dube syndrome diagnosed after spontaneous pneumothorax.

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The syndrome was reported for the first time in 1977 by Birt, Hogg, and Dube [1] who described small papular skins lesion distributed on the scalp, forehead, face, and neck in 15 of 70 members of the same family. The papula lesions develop after the age of 25 years, are autosomal dominantly inherited, and contain ectodermal and mesodermal components. Histologic examination of the lesions reveals fibrofolliculomas, trichodiscomas, and acrochordons. Subsequently patients may develop concomitant renal carcinoma and bullous lung diseases (eg, lung cysts or bullous emphysema), which are associated with recurrent pneumothorax.

A 43-year-old woman presented with a history of progressive dyspnea for 6 months. Chest roentgenogram revealed a left pneumothorax that was treated with drainage. Thoracic computed tomographic scan showed multiple bilateral cystic lesions of the lung (Fig 1). Family anamnesis revealed a Birt-Hogg-Dube syndrome (BHDS) in a member of her family. The patient indeed had papulous lesions on her face as described in the literature (Fig 2). Histologic examination of one of them revealed a fibrofolliculoma. The patient underwent a left thoracoscopy with wedge resections of the pulmonary cystic lesions for persistent pneumothorax (Fig 3). Histologic assessment revealed benign cysts and the absence of lymphangiioleiomyomatosis, which was one of the differential diagnosis revealed by computed tomographic scan images.

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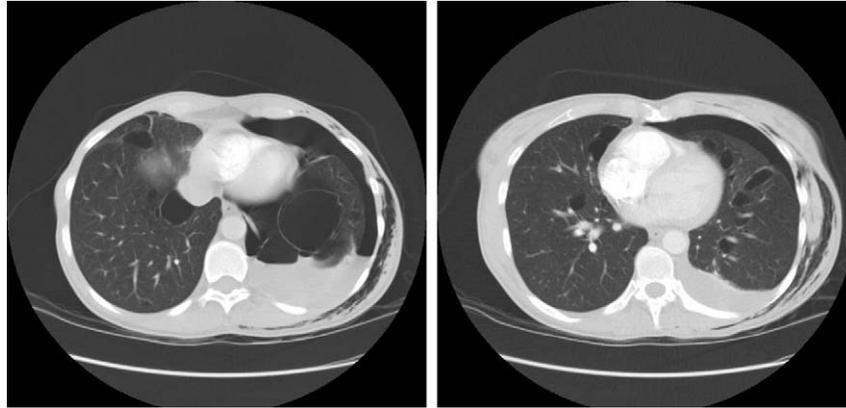


Fig 1. Computed tomographic scan images revealing bilateral cysts of the lung in the upper lobe (left) and lower lobe (right) associated with left pneumothorax.

Comment

Birt-Hogg-Dube syndrome describes a rare, autosomal dominant inherited pathology consisting of papular skin lesions (ie, fibrofolliculomas) on the face, neck, forehead, and scalp [1, 2].

Multiple or bilateral renal carcinomas, particularly chromophobe renal carcinoma, and renal oncocytomas have been reported in association with this syndrome [3-6]. Pulmonary cysts and spontaneous pneumothoraces have also been increasingly reported in association with BHDS [6]. Other associated symptoms have been described, such as large connective tissue nevus, parathyroid adenomas, flecked chorioretinopathy, bullous emphysema, lipomas, angioliipomas, parotid oncocytomas, and multiple oral mucosal papules [7]. Colonic polyps and colonic adenocarcinoma had been previously associated with BHDS; however, a recent large cohort study failed to demonstrate such a correlation [8]. Likewise medullary thyroid cancers have been found in 9 members of the original family described by Birt and colleagues [1], but not in subsequent cases.

Little is known about the pathophysiology of BHDS. The cause of mesodermal proliferation is unknown, but autosomal dominant inheritance has been identified in patients with BHDS. Recently it has been demonstrated that BHDS maps to chromosome 17p11.2 [4, 9].



Fig 2. Fibrofolliculomas on the face of the patient.



Fig 3. Intraoperative cystic pulmonary lesions of the left upper lobe (left) and the left lower lobe (right).

No specific medical treatment exists for the cutaneous lesions. The principle concern of BHDS is its association with renal carcinoma. Annual physical examinations and renal ultrasound screening should be proposed to those patients as well as to their relatives. Birt-Hogg-Dube syndrome is a rare entity that has to be suspected in cases of multiple pulmonary cystic lesions associated with papular skin lesion of the head and upper extremities.

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