follow-up, and computed tomography done at 11 months postoperatively showed an intact repair.

Comment

The diaphragm is formed by fusion of four structures: the pleuroperitoneal membranes; the septum transversum; the dorsal mesentery of the esophagus; and the lateral body walls [5]. Eighty percent of congenital defects involve the posterolateral diaphragm (Bochdalek gap). One percent to 6% of congenital defects are anterior and caused by failure of fusion of the septum transversum to form the pars sternalis portion of the diaphragm [3]. This specific entity has been described in Cantrell’s pentalogy of defects of the sternum, abdominal wall, pericardium, anterior diaphragm, and congenital heart disease (most commonly as tetralogy of Fallot) [2, 4]. Most hernias are discovered during maternal sonography or in the neonatal period owing to cardiorespiratory distress, often as a result of coexisting malformations. That explains the neonatal mortality rate of 40% to 50% [1–3, 6]. Our case is the 16th report of an isolated intrapericardial diaphragmatic hernia, and the third in an adult. Both open and laparoscopic techniques have been utilized in treatment but today the laparoscopic approach is preferred. For large defects, mesh repair that covers 2.5 cm to 3 cm beyond the defect should be effective.

The lessons to be learned are anterior congenital diaphragmatic hernias (1) result from failed development of the septum transversum of the pars sternalis of the anterior diaphragm; (2) may be initially asymptomatic and not present until later in life owing to incarceration; and (3) should be approached laparoscopically.

References


Diffuse Pulmonary Neuroendocrine Cell Hyperplasia Involving the Chest Wall

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Diffuse pulmonary neuroendocrine cell hyperplasia (DIPNECH) is characterized by a diffuse hypertrophy of neuroendocrine cells along the distal bronchioles. This condition is characterized by obstructive lung physiology and the development of small carcinoid tumors. We present a case of DIPNECH in a patient undergoing surgery for a primary lung adenocarcinoma. Interestingly, the chest wall also demonstrated involvement of DIPNECH indicated by the presence of small carcinoid tumors. The absence of any lung carcinoid tumor greater than 5 mm and the absence of lymph node metastases render the chest wall involvement unlikely to represent metastatic disease.


Pulmonary carcinoid tumors represent 1% to 2% of all lung malignancies and are pathologically classified as typical or atypical, depending on the degree of mitotic activity and the presence or absence of tumor necrosis. A recently described condition on the same spectrum of disease is diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH), which principally involves the distal airways and frequently is associated with numerous tiny carcinoid tumorlets, which are defined as lesions less than 5 mm.

We present a case of a patient who underwent surgical resection of a primary lung adenocarcinoma who was incidentally found to have DIPNECH. Interestingly, the...
parietal pleura also had carcinoid tumorlets, which is difficult to explain. An etiology related to de novo growth or metastases seems equally unlikely.

An 81-year-old woman with an extensive smoking history had a solitary spiculated and noncalciﬁed lesion in the right lower lobe (Fig 1). She underwent a transbronchial biopsy, and the pathology was consistent with adenocarcinoma. Computed tomography and positron emission tomography imaging did not show any evidence of metastatic disease. She had an acceptable performance status, and her pulmonary function test results suggest mild obstructive disease with a predicted forced expiratory volume - 1 second of 66%. She was scheduled for a video-assisted thoracic surgery (VATS) lobectomy, but she was found to have multiple nodules on the parietal pleura. Two of these nodules were biopsied, and the preliminary pathology conﬁrmed the presence of malignancy; therefore, the procedure was aborted. Upon further analysis of these specimens, it was clear that the histology of the small chest wall tumors was consistent with a typical carcinoid tumor rather than metastatic adenocarcinoma (Fig 2A). One week later, she returned for the planned VATS right lower lobectomy. The procedure was not complicated, and she was discharged home 3 days later. Her ﬁnal pathology revealed a 1.5-cm T1aN0 adenocarcinoma (Fig 2B). Incidentally, she was also noted to have multiple small carcinoid tumorlets in the lung parenchyma, consistent with a diagnosis of DIPNECH (Fig 2C). There was no evidence of adenocarcinoma or carcinoid tumor in any peribronchial or mediastinal lymph nodes. Adjuvant chemotherapy was not offered for her stage IA adenocarcinoma. She remained disease free for 15 months, but then developed distant metastases from the adenocarcinoma. There was no evidence of recurrent carcinoid disease.

**Comment**

Our growing understanding of the pathophysiology of DIPNECH indicates that it is clearly a primary pulmonary process. Neuroendocrine cell proliferation leads to local release of bombesin and gastrin-releasing hormone, which induce chemotaxis of ﬁbroblasts and subsequent bronchoconstriction [1]. Clinically, the most frequent symptoms noted are cough (71%), dyspnea (63%), and wheezing (25%). The vast majority of patients diagnosed with DIPNECH are middle-aged women. Radiographic features consistent with this diagnosis include ground-glass opacities, mosaic attenuation with air trapping, endobronchial wall thickening, and small pulmonary nodules [2].

A surgical biopsy is frequently required to conﬁrm the diagnosis of DIPNECH, although the diagnosis can be conﬁrmed in a minority of cases with a transbronchial biopsy. The histologic features of DIPNECH are proliferation of neuroendocrine cells along the bronchioles that is initially conﬁned to the mucosa. When the proliferation extends transmurally into the lung parenchyma, the nodule meets the deﬁnition of a carcinoid...
tumorlet. When a tumorlet exceeds 5 mm, the lesion is defined as a carcinoid tumor. Interestingly, a review of surgical specimens of peripheral carcinoid tumors revealed that 19 of 25 (76%) also demonstrated histologic evidence of neuroendocrine cell hyperplasia surrounding the tumor [3]. Eight patients also demonstrated evidence of constrictive bronchiolitis. These histological observations support the hypothesis that DIPNECH and carcinoid tumors are on the same spectrum of disease.

Management of patients with DIPNECH remains controversial due to the relative novelty of this diagnosis, as well as the indolent and diffuse nature of the disease. Most centers recommend serial imaging for the majority of patients and surgical resection for lesions exceeding 10 mm in diameter. The greatest risk of DIPNECH relates to bronchial obstruction rather than the malignancy. Chemotherapeutic treatment for the carcinoid tumorlets or DIPNECH is currently not supported, but this may be related, in part, to the paucity of patients with this condition diagnosed. In addition, there is limited information indicating progressive pulmonary fibrosis and obstructive disease despite treatment with cytotoxic agents. Steroids can improve pulmonary function and limit long-term progressive pulmonary fibrosis [2].

The most interesting point of the case presented here is the unresolved issue regarding the nature of the chest wall carcinoid tumors and whether they represent metastatic disease or tumorlets arising de novo on the parietal pleura. The pattern of disease progression associated with carcinoid tumors is essentially the same as other types of non–small cell lung cancer where mediastinal lymph nodes are the most likely location for early metastatic disease. In the current patient, all the mediastinal lymph nodes were negative for metastases from the carcinoid tumor and the adenocarcinoma. This finding increases the likelihood that the pleural-based tumors represented de novo carcinoid tumors arising directly from the chest wall. The current literature contains a few examples of carcinoid tumors that metastasized to the chest wall, but these cases were associated with large tumors, and the chest wall metastases developed up to 20 years after the primary tumor was treated surgically [4,5]. The limited disease, the absence of any carcinoid tumor greater than 5 mm, and the absence of lymph node metastases reduces the probability that the chest wall tumors were metastases.

Conversely, carcinoid tumors originate from amine precursor uptake and deamination (APUD) cells that are located throughout the respiratory and gastrointestinal systems. APUD cells have not been described previously on the chest wall, rendering the de novo origin hypothesis improbable.

Our decision against adjuvant chemotherapy for a surgically resected T1N0 adenocarcinoma is consistent with the standard of care. The unfortunate development of distant metastases, which will likely lead to the patient’s demise, is a known reality for some patients with lung cancer. The diagnosis if DIPNECH or the chest wall involvement of carcinoid tumorlets will likely have no meaningful effect on her long-term outcome.

References

Primary Mediastinal Hemangiopericytoma Treated With Preoperative Embolization and Surgery
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Hemangiopericytomas are rare tumors originating from vascular pericytes. The mediastinum is an extremely uncommon site with only a few cases reported. Diagnosis is based on histopathology and immunohistochemistry, which differentiates them from synovial sarcoma and solitary fibrous histiocytoma. They have a variable malignant potential. Treatment is mainly surgical extirpation as the role of adjuvant therapy is controversial. Preoperative embolization has been sparingly used. We report a case of primary mediastinal hemangiopericytoma in a 47-year-old man treated successfully with preoperative embolization and surgery.


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