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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mediastinal HPC managed successfully by preoperative embolization and surgery.

A 47-year-old man came to our institution with complaints of dyspnea on exertion and dry cough for the previous 3 months. Physical examination was essentially normal. Laboratory studies were within normal limits. A chest roentgenogram showed a grossly widened mediastinum without effusion or lung parenchymal abnormalities. A contrast-enhanced computed tomography (CT) scan of the chest revealed a 18.3 × 9.3 × 10.5 cm soft tissue mass in the middle and posterior mediastinum in the midline. The mass extended from the fourth to the 12th thoracic vertebral level with close proximity to the esophagus, carina, and descending aorta (Fig 1). On contrast administration, the mass demonstrated intense heterogenous vascularity with areas of necrosis and calcification (Fig 1). Additional work-up in the form of a whole-body positron emission tomography/CT scan was negative for regional or distant metastases. The tumor markers alpha-fetoprotein, carcinoembryonic antigen, CA 19-9, neuron-specific enolase, and squamous cell carcinoma antigen were within normal limits.

Based on the highly vascular nature of the tumor on imaging, a preoperative embolization was performed through the femoral approach. During the procedure, the tumor was found to be supplied by five major feeders, namely, the inferior phrenic, left gastric, and three bronchial arteries (Fig 2). All of these were successfully occluded using 300 to 700 micron-sized emboli. The surgery was done 36 hours later.

The patient underwent right posterolateral thoracotomy through the fifth intercostal space. During the operation, the mass appeared to be mediastinal in origin. The esophageal part of the dissection was completed from there. Thereafter, a left posterolateral thoracotomy was done, and the aortic and pericardial dissection was completed. A sliver of lung tissue was taken on both sides for margin. The mass measured 18 × 9 × 11 cm and weighed 900 g. Grossly, the resected specimen was a lobulated encapsulated mass with focal hemorrhages (Fig 3). The cut surface was smooth, elastic, and pale brown. Microscopic examination showed round and spindle cells surrounded by thin-walled, endothelium-lined vascular channels, giving a “staghorn” appearance to the vessels as typically seen in HPC (Fig 4). There were no mitotic figures. The lung tissue taken as margins was uninvolved by the tumor, confirming R0 resection. The tumor cells were immunoreactive only for CD34 (Fig 5). The postoperative course was uneventful. No adjuvant therapy was given because of the absence of high-risk features. The patient is being followed up with contrast-enhanced CT scan of the chest every 3 months. He is alive and disease free 21 months after the operation.

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**Fig 1.** Contrast-enhanced computed tomography chest showing the large vascular mediastinal mass displacing the esophagus. Areas of necrosis and calcification are visible.

**Fig 2.** Preoperative angiography showing the feeder vessels to the tumor: (A) first bronchial; (B) second bronchial; (C) inferior phrenic; and (D) left gastric.
Comment

Hemangiopericytoma is a potentially malignant, uncommon tumor arising from pericytes in the small vessels. In the thorax, these are pericytes that surround the basement membrane of capillaries and small venules within the lung parenchyma [2]. Our case was an intrathoracic mediastinal HPC, which is extremely rare. Only a few case reports are available in the literature [4]. The pulmonary variety is more common. There are no diagnostic clinical or radiographic features and mostly presents as an asymptomatic, noncalcified solitary mass on chest roentgenogram. They are composed of closely packed spindle cells and prominent vascular channels. The histologic differential diagnosis mainly includes solitary fibrous tumor and the synovial sarcoma [2]. However, unlike HPC, synovial sarcomas are often immunoreactive for both keratins and epithelial membrane antigen [3]. There is no single feature, including histologic type or DNA ploidy, that predicts biologic aggressiveness [2]. Malignant HPC is recognized by its increased mitotic rate, tumor size, and foci of hemorrhage and necrosis [2].

The preoperative diagnosis remains a concern. When a mass appears to be radiologically resectable, a thoracotomy is often performed without histologic diagnosis. Others have attempted to obtain a preoperative diagnosis even in tumors that are clearly resectable if high vascularization is suspected on imaging techniques [2]. Surgical radical excision is the treatment of choice for HPC, although the criteria for determining the area of resection have not been established. Hansen and coworkers [5] believed it necessary to consider all HPCs as malignant and perform extended surgery. Chemotherapy or radiotherapy have been recommended in the adjuvant setting but is considered to be almost ineffective [2], although Rusch and colleagues [6] reported that combination or single therapy with adriamycin was effective against metastases. Jalal and colleagues [7] reported that preoperative radiotherapy of large chest wall HPCs significantly reduced the vascularity of the tumor and made complete resection much easier. Morandi and associates [8] recommended preoperative percutaneous embolization of hypervascular mediastinal tumors, to allow a safe, complete removal of the lesion later.

The 5-year survival of patients with any organ HPC has been reported to be 85%, whereas that of patients with a tumor of pulmonary origin is 30% to 35%. Approximately 50% recur within 5 years [2, 5]. Distant metastases to liver, brain, and bone have also been reported [5].

In conclusion, HPC is an uncommon variety of soft tissue tumor, mediastinum being an extremely rare site. Diagnosis is based on histopathologic evidence of “staghorn” arrangement of vessels and immunohistochemistry. The biologic behavior of these tumors is variable.

Fig 3. Gross photograph of the resected tumor showing well-encapsulated bosselated appearance.

Fig 4. Photomicrograph showing the classical “staghorn” appearance with intense vascularity. (Hematoxylin and eosin stain, original magnification ×10).

Fig 5. Immunohistochemical photomicrograph showing CD34 positivity (magnification ×100).
and is determined by the presence of mitosis, size, necrosis, and hemorrhage. Surgical resection, although technically difficult, is the treatment, although the extent of resection remains controversial. Preoperative embolization may be of benefit for radiologically vascular tumors, as in our case.

References

The Use of Extracorporeal Membrane Oxygenation Therapy in the Delayed Surgical Repair of a Tracheal Injury
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Acute tracheal injury secondary to intubation can present with varying degrees of severity. Onset of symptoms occur hours or even days after the initial injury. A 34-year-old woman required surgery for a large tracheal tear after emergency intubation. The inability to adequately ventilate combined with secondary aspiration injury required that the patient be placed on extracorporeal membrane oxygenation before undergoing surgery. This case demonstrates the use of extracorporeal membrane oxygenation to manage a patient awaiting surgery for severe tracheal tears.


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A cute tracheal injury due to intubation is quite rare, with a reported 1 in 20,000 single-lumen intubations resulting in injury, 15% of which occur in an emergency setting [1]. The resulting tear can be small, managed conservatively: or severe, requiring emergent surgery, increasing the risk of morbidity and mortality.