Case report

Serial transarterial embolization for the management of unresectable malignant pulmonary hemangiopericytoma: A case report and review of the literature

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ABSTRACT

Hemangiopericytomas (HPC) are perivascular neoplasms that are rarely encountered as primary lung malignancy. Surgical resection remains the mainstay therapy of HPC. A 37-year old African American female initially presented with fatigue, chest pain and palpitations. Chest radiography showed a well-circumscribed large heterogeneous vascular mass of the right hemithorax. She underwent a CT-guided biopsy of the thoracic mass and was diagnosed with low-grade pulmonary hemangiopericytoma. High tumor vascularity burden and liver metastases precluded her from being a surgical candidate. Three years ago, she referred to our facility seeking further management. She did not tolerate systemic chemotherapy. Alternatively, she had been successfully managed with serial transarterial embolization and oral angiogenesis inhibitor to date. This therapeutic approach can be further explored for long term control of unresectable pulmonary HPC.

1. Introduction

Hemangiopericytomas (HPC) are rare vascular neoplasms arising from the pericytes that surround the capillary blood vessels and are rarely of thoracic origin [1,2]. Surgical excision is the standard of care as HPC have a high recurrence rate as well as 20–30% of the cases may show a malignant course [1]. Chemotherapy for unresectable and disseminated HPC can lead to complete or partial remission of the disease in 50% of the cases [1]. Other therapeutic modalities are needed. Preoperative transarterial embolization (TAE) was reported in surgical treatment of HPC [8]. We present a case of unresectable pulmonary HPC that has been successfully managed with serial TAE and oral angiogenesis inhibitor.

1.1. Case description

A 37-year-old African American female presented to our cancer center three years ago for evaluation of therapeutic options of unresectable pulmonary HPC. She has been complaining of profound fatigue, right-sided chest pain, non-productive cough, unintentional 20 pound weight loss and night sweats. Her only previous medical history was an incidental heart murmur detected in childhood. Physical exam was unremarkable with the exception of absent breath sounds over the lower third of the right hemithorax. Diagnosis was previously made with CT guided lung biopsy of the thoracic mass. Histopathology revealed spindle cell neoplasm with occasional mitoses which is characteristic of solitary fibrous tumor/hemangiopericytoma. Immunohistochemical markers were strongly positive for CD34, CD99, and bcl2 and negative for keratin, CD31, factor VIII, desmin, S-100 and melanA, consistent with the diagnosis of hemangiopericytoma. Contrast enhanced CT scan of the chest, abdomen and pelvis revealed a large 12.4 x 14.0 x 12.5 cm heterogeneous mass on the anterior inferior aspect of the right hemithorax, numerous draining veins into the left atrium, and leftward deviation of the mediastinum caused by mass effect (Fig. 1-A,B). There were no signs of any destructive changes to the surrounding ribs. The liver demonstrated numerous enhancing lesions, the largest measuring 5.6 x 4.6 cm in the left hepatic lobe (Fig. 1-D). CT angiography showed feeder vessels from the inferior phrenic branch and intercostal vessels. Additional hypervascular masses were found within the left and right hepatic lobes. Positron emission tomography (PET) obtained showed no evidence of fluorodeoxyglucose (FDG) avid malignancy.

After multidisciplinary discussion with medical, radiation and surgical oncology, the tumor was deemed inoperable in view of its extensive vascular burden, risk of catastrophic bleed and spread to the liver. A decision to initiate palliative chemotherapy has been determined. The patient did not tolerate a chemotherapy regimen of temozolomide and bevacizumab due to intractable nausea and vomiting.

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To offer local tumour control, the patient underwent embolization of the thoracic tumour feeding vessels which were identified on arterial phase CT images as hypertrophied arteries. There was concern for non-target embolization with embolizing these large blood vessels. Prior to embolization, each major tumor feeding vessel in the liver and chest was assessed to confirm that there was no large shunt. Overall, although there were massive draining veins, there did not seem to be any direct artery to vein shunts that were not avoidable. Initial TAE was successfully performed on the left gastric artery and right inferior phrenic artery as well as the segment III hepatic artery. Embolics used were 40-120, 300-500, 500-700, and 700-900 micron Embospheres (Merit Medical Systems, Utah, USA). These were given from small to large sequentially until approximately five heart beat stasis. Although unlikely that the pulmonary artery would feed this thoracic tumor anyway, pulmonary supply in fact was confirmed to not be present on cross sectional imaging. As expected, recovery was complicated by post-embolization syndrome (PES) that required hospitalization for intractable nausea and vomiting. Two months later, CT imaging of the thorax and abdomen revealed successful partial necrosis with areas of necrosis causing mass effect and leftward deviation of the mediastinum. Therefore, decision has been made to initiate an oral angiogenesis inhibitor sunitinib. Patient elected for a one-year surveillance interval. Subsequently, PET scan was performed to monitor disease progression. The thoracic mass showed a curvilinear focus of mildly increased FDG uptake along the superior lateral aspect with no real significant change of size. FDG avid, low attenuation of the left hepatic lobe lesions were demonstrated. Clinically, the patient was thought to have disease progression and underwent the fourth TAE. During the procedure, successful bland embolization to segments IVa and IVb left hepatic artery branches, the right hepatic artery, a branch of the inferior phrenic artery, the inferior epigastric artery and the right internal thoracic artery (Fig. 2-B) followed by Gelfoam embolization were performed. Through sonographic guidance, the right fifth intercostal artery was entered, followed by bland and Gelfoam embolization on three different large tumor feeder distributions. Post-procedure, the patient developed...
hemopneumothorax that was managed by short term chest tube. To date, the patient is being followed by thoracic oncology.

2. Discussion

2.1. Definition

Hemangiopericytomas represent a subtype of rare vascular tumors arising from pericytes surrounding the capillaries. First described by Stout and Murray as Zimmerman’s pericytes, these modified smooth muscle cells act to regulate luminal patency [3]. Location varies, but most tumors arise within the head and neck, muscles of the extremities and within the retroperitoneum [1,2]. Thoracic origins represent only 10% of documented cases, but have been reported to originate within the mediastinum, trachea and lungs [2].

2.2. Symptomatology

Symptoms vary depending on the type and degree of organ involvement. Primary pulmonary HPC are usually diagnosed incidentally on chest radiography unless large enough to cause symptoms of chest pain, cough, dyspnea or hemoptysis [4,5]. Other reported symptoms found associated with pulmonary solitary fibrous tumor and HPC are night sweats, hypoglycemia due to the production of insulin-like growth factor and pulmonary osteoarthropathy [1,4].

2.3. Diagnostic features and imaging

There are no reported diagnostic features on chest radiography or CT to confirm a diagnosis of HPC. Chest radiography usually demonstrates a sharply outlined mass while contrast enhanced CT reveals a heterogeneous mass with areas of necrosis and/or calcifications [4]. Pulmonary HPC can be differentiated from other pulmonary lesions by the absence of bronchial communication and the presence of hemorrhage, necrosis and a reticulin network [6]. On histopathology, these lesions demonstrate a staghorn appearance composed of vessels with tightly packed spindle cells [2,4]. Antibody staining against vimentin and collagen type IV are positive, while negative for S-100, desmin, laminin, cytookeratins, and factor VIII-related antigen [4,5]. The immunohistochemical expression of vascular endothelial growth factor receptor (VEGFR) has been described in the tumoral cells of HPC and in the endothelium suggesting that the autocrine and paracrine activation of the VEGF-VEGFR pathway may participate in the biology of HPC [9].

2.4. Criteria for malignancy and recurrence

Literature varies in regards to the criteria for malignancy. Those that support the conventional method recognize high mitotic rates, size and foci of hemorrhage and necrosis to be diagnostic for malignancy, while others define malignancy based on tumor tendency to reoccur and spread [2,7]. Other criteria have been proposed to better define malignancy in tumors originating within the thorax including lesions that invade the chest wall or mediastinum, angiolymphatic invasion and recurrent or metastatic disease [4]. Reoccurrence of the lesions is as high as 50% and varies based upon tumor location [7]. Those with pulmonary origin tend to reoccur at rates of 30–35%, and those that do reoccur, 80% will be present within the first year [7]. Metastatic disease occurs within the liver, brain and bone and the 5-year survival of patients exhibiting primary pulmonary origin is between 30 and 35% [2].

2.5. Modalities of management

Due to their unpredictable nature and high recurrence rates especially within the first year, pulmonary HPC are treated aggressively, with the treatment of choice being surgical resection and close long term follow up [2,4–7]. More recently, TAE through either chemical or mechanical means has gained popularity in the pre-operative interval to limit intraoperative bleeding and to decrease the risk of catastrophic bleeding events of highly vascularized lesions [2,8]. Recommendations have now been made to incorporate the use of preoperative angiographic studies to detect tumor feeder vessels prior to surgical resection [8,9]. Radiotherapy has been cited as another potential adjunct to surgery to facilitate better surgical outcomes [2]. However, HPC are relatively radiosensitive [1]. Chemotherapy has shown some efficacy in metastatic disease. Adriamycin alone or in combination with other chemotherapy agents have been associated with complete or partial remission of disease in up to 50% of patients [1,11]. The efficacy of bevacizumab and temozolomide in metastatic HPC has been described in a population-based analysis with an overall response rate of 21.4%, and a median progression free survival of 17 months as previously reported [10]. Sunitinib, a tyrosine kinase inhibitor of VEGFR, PDGFR and fibroblast growth factor, along with other angiogenesis inhibitors have been used in case reports of metastatic HPC and showed response to therapy and disease stabilization [11].

3. Conclusion

There are limited therapeutic options for unresectable pulmonary HPC. This case highlights the potential role of serial TAE combined with an angiogenesis inhibitor in long term control of this disease. Further trials are needed to explore whether this approach is effective for control of local tumor and disseminated disease.

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