Oncology and Cancer Case Reports

El-Hasbani and Assaker, Oncol Cancer Case Rep 2018,4:2

Case Report Open Access

Epithelioid Hemangioendothelioma Involving the Left Apical Lung, Lingula and Right Atrium

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Abstract

Epitheloid hemangioendothelioma is a tumor of vascular origin that could involve different organs such as the heart, lungs, and liver. In this case report, we present a 62-year-old patient who presented for one-month history of dry cough and hoarseness. A CT scan of the chest showed multiple scattered bilateral pulmonary nodules. A CT guided biopsy of the nodules was done to be consistent with the diagnosis of epitheloid hemangioendothelioma positive for CD 31 and CD 34 involving the right atrium, left apical lung, and lingula. A subsequent excision of the tumor was done. According to our literature review, this is the first reported case of epitheloid hemangioendothelioma to have this unique involvement of right atrium, left apical lung, and lingula. Additionally, the presentation of the patient is unique in terms of presentation where no cardiac symptoms or severe respiratory symptoms were noted. The diagnosis of epitheloid hemangioendothelioma tumor necessitates imaging, cytology, and pathology. The treatment is excision with no standard recommendation for the treatment with chemotherapy post-operatively.

Keywords: Epitheloid hemangioendothelioma; Cardiac tumors; Lung tumors; Tumor involving the right atrium left apical lung and lingula

Introduction

Epitheloid hemangioendothelioma (EHE) is an uncommon tumor of vascular origin. It has been primarily seen in soft tissues, but also commonly reported to appear in several locations such as lung, liver, and bone [1,2]. Although previously considered as a borderline vascular lesion, it is now under the classification malignant tumors due to its locally aggressive behavior and ability to metastasize. Etiology and associated risk factors are still unknown. Herein, we report a case of a 62-year-old male presenting with EHE involving the left apical lung, lingula, and right atrium which is the first to be reported in the literature in terms of distribution.

Case Presentation

A 62-year-old Caucasian male known to have hypertension and diabetes mellitus type 2 presenting with a one-month history of dry cough associated with hoarseness. He had no hemoptysis, wheezes, chest pain, or dyspnea. Also, he had no night sweats, weight loss, fatigue, loss of appetite, or low-grade fever. The patient wasn't positive for HIV throughout his life.

The patient's surgical history is positive for laminectomy. His hypertension was controlled on Bisoprolol 5 mg once daily. He didn't have any allergies to food or drugs. He wasn't an intravenous (IV) drug user. He didn't have any travel history. However, the patient reported that he's a smoker (20 pack year). Review of systems was negative.

On physical exam, he was 160 cm in height and 92 kg in weight. His vital signs were within acceptable limits. The respiratory exam revealed good bilateral air entry with fine rhonchi. Cardiac and abdominal exams were normal.

A primary chest x-ray revealed a well-defined 7 × 8 mm nodular opacity in the projection of the right middle lung field; the heart was borderline in size (Figure 1). A CT scan of the chest showed multiple scattered bilateral pulmonary nodules predominantly in a peripheral and perilymphatic distribution, all of which showing irregular borders (Figure 2). The nodules appeared more numerous at the postero-basal segments of the lower lobes. There was no evidence of pleural or pericardial effusion. Following up with a CT guided biopsy, the alveolar parenchyma was partially replaced by a sclerotic fibrosis that contained a proliferation of giant cells groups in cords with foci of tumor necrosis. The proliferation was pleomorphic associating small cubic cells with a pale cytoplasm to giant cells with eosinophils abundant cytoplasm containing a hollow cavity. The biopsies had a high expression of CK7 with positive vascular markers CD31 and CD34, as well as focal markings for CKC. After further workup, the tumors were found to be involving the right atrium at the junction of superior vena cava (SVC), as well as the apex of the left lung and lingula.

A right atriotomy was made. A large tumor of the right atrium was found with a large implantation base about 2 cm of diameter reaching the atrial septum and inferior vena cava (IVC). All vegetations were removed prior to the removal of the right atrial tumor with its base including the full thickness of the right atrium (RA) cause a gap in the right atrium and the IVC. Consequently, the IVC was repaired as well the RA.

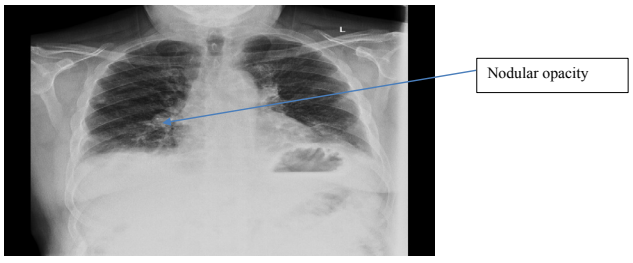

Figure 1: Nodular opacity in the projection of the right middle lung field.

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Received: July 18, 2018; **Accepted:** July 31, 2018; **Published**: August 03, 2018

Citation: El-Hasbani G, Assaker R (2018) Epithelioid Hemangioendothelioma Involving the Left Apical Lung, Lingula and Right Atrium. Oncol Cancer Case Rep 4: 148.

Tumor was sent to pathology and found to be multifocal in the apical region of the lung, lingula, and right atrium consistent morphologically and immunophenotypically with the diagnosis of epitheloid hemangioendothelioma (Figure 3). The tumor was composed of spindle shaped cells and more frequently of large polygonal cells with eosinophillic ground glass cytoplasm, pleomorphic nuclei sometimes nucleated with monstrous figures, frequently arranged around central necrosis. Also, some areas showed epitheloid cells set within a fibromyxoid stroma, and other areas showed pseuotrabecular pattern. Within the lung, the tumor was infiltrating extensively the pleura and showed numerous sub pleural carcinomatosis (Figure 4). The atrial tumor was infiltrating the pericardium and showed multifocally pseudo papillary pattern and extensive neurosis with large calcified area occupying the large part of the tumor. The tumors were extensively positive by immunostain with CD31 and CD34 (Figures 5 and 6) that was consistent with the above diagnosis. Moreover, it was CK 5/6, p63, and TTF1 negative, ruling out squamous cell carcinoma and adenocarcinoma of the lung.

Patient had a smooth postoperative course with no signs of complications and was discharged on pain medications. He didn't receive any adjuvant chemotherapy.

Discussion

Epitheloid hemangioendotheliomas (EHE) are rare malignant tumors of vascular origin that arise mostly in soft tissues and visceral organs and, less commonly, large veins. EHE has a predilection for women thought to be because of the participation of estrogen receptor in the proliferation of the tumor [3]. On the other hand, the patient presented in this case report is a male which contributes to the uniqueness of the case.

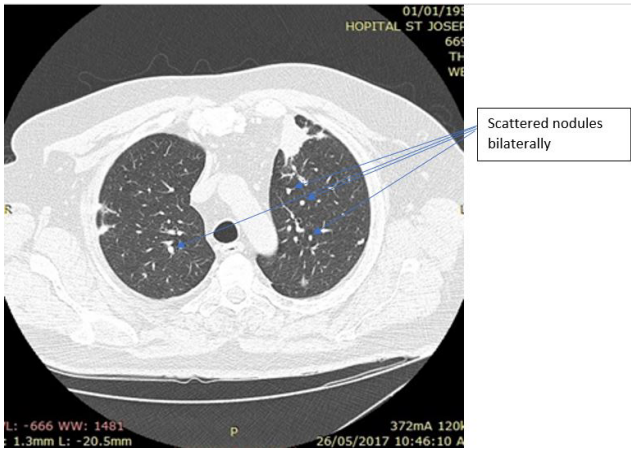

Figure 2: Multiple scattered bilateral pulmonary.

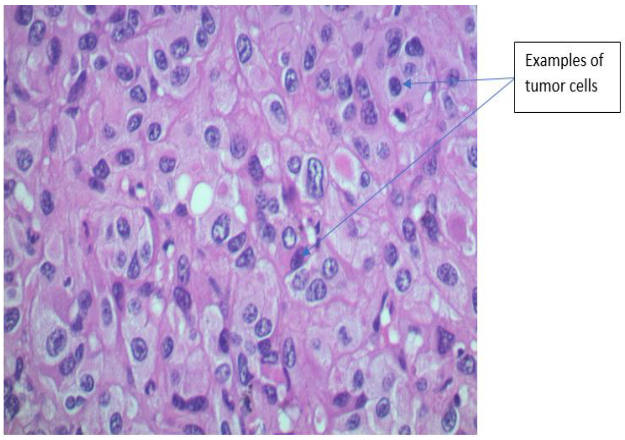

Figure 3: Tumor cells of vascular origin.

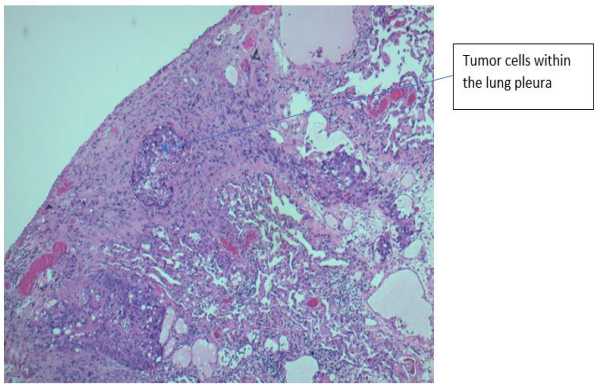

Figure 4: Tumor cells infiltrating the lung pleura.

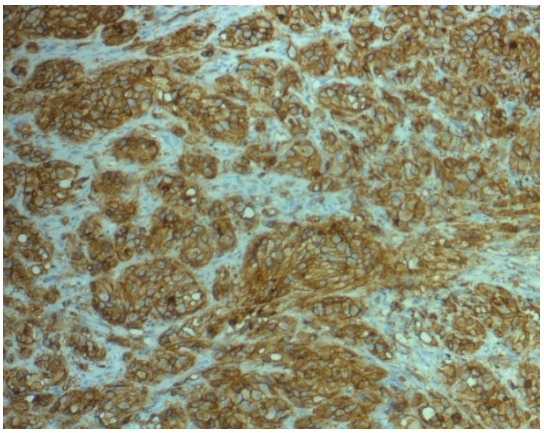

Figure 5: CD31 stain.

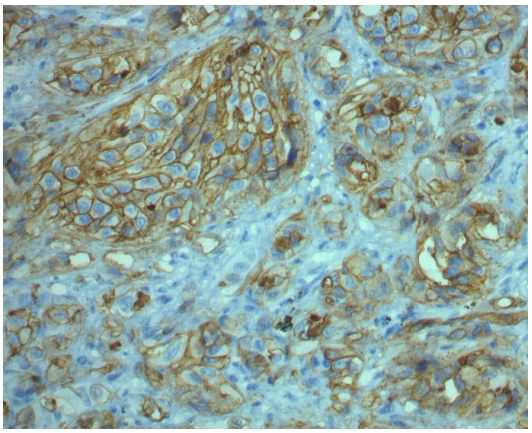

Figure 6: CD34 stain.

Arising from a vessel which is usually a medium-sized or large vein in the lung, liver, bone, and soft tissues, EHE can result in a variety of symptoms depending on the organ of involvement [4]. When EHE involves multiple organs simultaneously such as the heart and lungs, it may be difficult to determine if the tumor is multicentric/multifocal from the beginning or there is a primary lesion at one site with metastases to the other tissues [5].

Based on the literature review, there are 4 cases of EHE [6-9] arising in the right atrium which is the most common site for cardiac EHE. These tumors can involve nearby structures such as SVC or lungs as what occurred in our case because they have an unpredictable outcome and may proliferate indefinitely [10]. There have been around 100 cases of pulmonary EHE reported in the literature [11-14]. To our knowledge, this is the first reported case of co-occurrence of EHE in the left apical lung and lingula with pleural involvement, and right atrium.

A cardiac EHE can result in heart failure caused by tumor obstruction of cardiac chambers or valves resulting in cardiac and respiratory symptoms. The patient presented for only a history of dry cough and hoarseness which is special for a cardiac involvement of EHE.

However, patients having cardiac EHE with SVC involvement can often be asymptomatic despite the evidence of occlusive disease confirmed on venography. Such a tumor is often incidentally discovered after an abnormal chest radiograph [2] which matches with our case where the tumor was suspected on the chest x-ray after a vague clinical presentation. The EHE tumors range from discrete nodules to a mass of coalesced nodules. Radiographically, contrast-enhanced CT reveals a well-defined rounded mass of fat attenuation occluding the lumen of the SVC in case of SVC involvement [15].

Pathologically, the tumors are composed of short stands or solid nests of rounded to slightly spindled endothelial cells, characterized by an abundant eosinophilic cytoplasm and large hyperchromatic nuclei [16]. In the presented case, the tumor had large polygonal cells with eosinophilic ground glass cytoplasm and pleomorphic nuclei which matches the pathological presentation of EHE mentioned in literature.

Immunohistochemically, the right atrial tumor in the presented case was positive for CD 31 and CD 34, which are endothelial cell markers consistent with EHE. It was negative for TTF1, CK 5/6, and P63, which rules out squamous cell or adenocarcinoma of the lung. EHE usually expresses at least one endothelial marker such as CD31, CD34, factor VIII, and is mostly negative for cytokeratin. CD31 is highly sensitive and is a specific marker of endothelial differentiation [17] and was the most positive immuno-stain used for detecting tumor cells in our case.

Wide local excision is the treatment of choice for benign lesions. As for malignant lesions, they are best treated by radical excision because they tend to metastasize to lymph nodes and lungs [18]. Sometimes, remissions with chemotherapy have been reported [19]. There is no standard choice for chemotherapy for EHE. The choice of therapy depends on the malignant potential of the tumor, and the presence of metastasis at the time of diagnosis or pathological features. However, there are no recommendations for adjuvant therapy [9]. In our case, a wide local resection was made with removal of tumor vegetations. The patient didn't receive any adjuvant or neo-adjuvant chemotherapy.

A pulmonary EHE involving the pleura is considered aggressive with most patients surviving less than a year [20]. Patients having cardiac involvement of EHE have a favorable outcome after surgical resection even with rare pulmonary metastasis [9,21-23]. Our patient was symptom free at 1 month after the surgery. However, his clinical outcome for further duration needs to be assessed.

Conclusion

In conclusion, we present a case of a 62-year-old man who presented with a dry cough and hoarseness found to have high expressivity for CD 31 and CD 34 on CT guided biopsy. After a mass excision, the tumor was found to be an epitheloid hemagioendothelioma involving the right atrium, lingula, and left lung apex which is the first case to be reported in the literature involving all of these tissues. The patient had a good post-op response with no chemotherapy administered.

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