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Case Report
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Pulmonary Epithelioid Hemangioendothelioma: An Unusual Case Treated with Radiotherapy

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Abstract

Epithelioid Hemangioendothelioma (EHE) is a rare vascular neoplasm. Initially described as an intravascular bronchioalveolar tumor by Dail et al., it is derived from vascular endothelial and pre-endothelial cells [1]. It is classified as a malignant neoplasm, with slow growth and an indolent course [2]. It may present at different sites, however, in the lung, the tumor is usually bilateral, usually affects young people, and is more commonly seen in women [3]. In this case report we describe the case of an elderly woman with pulmonary EHE treated with surgical resection and radiotherapy. We review the relevant literature and explore the therapeutic potential of radiotherapy to prevent local disease recurrence. Written informed consent for the case to be published, inclusive of figures, case presentation and data, was obtained from the patient.

Case Presentation

This is a case of a 69-year-old female with a history of osteoarthritis and hypertension who presented with shortness of breath on exertion that had been steadily increasing in intensity over a period of four months and a non-productive cough for two months. Her chronic illnesses were controlled on enalapril, hydrochlorothiazide, and simvastatin. She was a lifetime non-smoker with occasional alcohol use and had no personal or family history of cancer. Physical examination was unremarkable and laboratory tests were all within normal limits. Chest radiography revealed a right paratracheal soft tissue density and subsequent Computed Tomography (CT) scan described a right paratracheal mass with a focus of calcification consistent with a nodal mass (Figure 1).

In order to further evaluate the mass, she underwent bronchoscopy and right anteroposterior thoracotomy with excision of the nodal mass adherent to superior vena cava and azygos vein. The initial pathological diagnosis suggested features consistent with metastatic osteogenic sarcoma; however, specialist consultation and immunohistochemistry revealed that it was, in fact, an epithelioid hemangioendothelioma associated with myelolipomatous changes and mature bone (Figure 2).

Immunohistochemistry showed tumor positive for CD31, Factor VIII, CD34 and negative for H-caldesmon, keratin, P63, S100 protein, desmin, myeloperoxidase, Glyc-c, CD45 and D2-40. A CT scan of the chest done a month later revealed a right apical pleural based lesion and posterior basal pleural based thickening with right medial basal consolidation and small right paratracheal

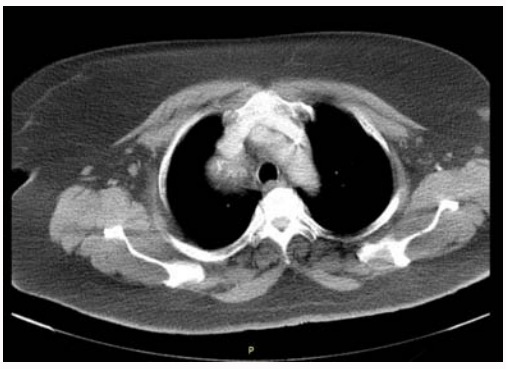

Figure 1: Axial CT scan image of the chest shows a right paratracheal soft tissue density with foci of calcification.

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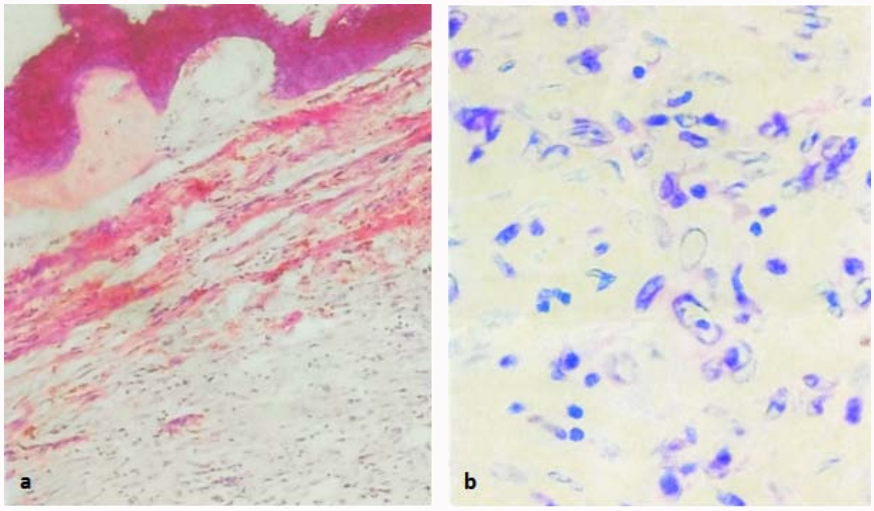

Figure 2: Sections showed tissue composed of elongated spindle cells with markedly pleomorphic nuclei and associated with myelolipomatous changes. In addition, bone and osteoid formations were seen with areas of tumor necrosis present. Low power (x10) H&E (a) and High power (x40) H&E (b).

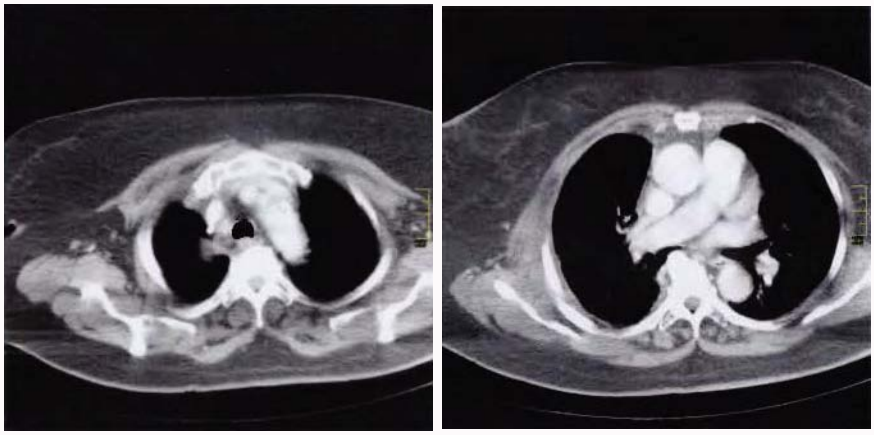

Figure 3: Axial CT scan images of the chest shows **(a)** a right medial apical pleural-based soft tissue mass and **(b)** a right posterior basal pleural thickening.

nodes (Figure 3). The patient received no further therapy following surgery by mutual agreement with her oncologist, and serial CT scans were recommended. Two years after diagnosis, CT chest described an increase in the size of the right posterior basal pleural based mass and a soft tissue lung mass extending from it measuring 3.3 cm × 1.4 cm × 2.2 cm (Figure 4). The patient was also noted to be symptomatic of right-sided chest discomfort at that time. A decision was made to proceed to radiotherapy. The patient was treated on a linear accelerator with the Three-Dimensional Conformal Radiation Therapy (3D-CRT) technique using 6MV photons. She received 64 Gy in 32F over the course of 6.5 weeks. Treatment was well tolerated with minimal adverse effects.

Since completion of treatment she has been followed up with serial CT scans every 6 months; with no evidence of relapsed or recurrent disease. Her clinical course has remained uneventful, now seven years since her original diagnosis.

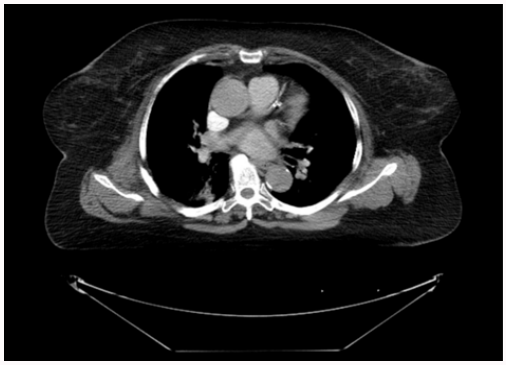

Figure 4: Axial CT scan image of the chest shows right posterior basal pleural-based thickening increased in size (0.8 cm × 2.1 cm × 2.7 cm) and with a soft tissue lung mass measuring 3.3 cm × 1.4 cm × 2.2 cm now extending from it.

Discussion

Epithelioid Hemangioendothelioma (EHE) is a rare vascular tumor. It was characterized initially by Dail and Liebow in 1975 as an aggressive form of a bronchoalveolar cell carcinoma but subsequently renamed to its current description in 1982 by Weiss and Enzinger due to its overlapping features between a hemangioma and an angiosarcoma [1,4]. In the most recent edition of the World Health Organization classification of sarcomas, EHE is defined as an independent disease from other vascular tumors and shows low- to intermediate-grade malignancy [2]. It can present at numerous sites with the most commonly involved organs being liver, lung and

bone [5]. Pulmonary Epithelioid Hemangioendothelioma (P-EHE) is usually an incidental finding as approximately 50% to 76% of patients are asymptomatic. Otherwise, symptoms may include cough and dyspnea as presented in this case; with other reports describing malaise, anemia and hemoptysis in some patients [6]. In a review of 248 case reports it was found that P-EHE was more common in women than men at a 1:4 ratio and a median age of onset at 36 years old; with a range from 7 to 83 years old [3]. Most patients present with bilateral nodular opacities on chest radiographs, showing little or no growth on serial CT scans; usually being less than 2 cm in diameter. Calcification as seen in this case is uncommon but may represent areas of ossified necrosis [7]. Poor prognostic factors include symptomatic patients, age ≥ 55 years old, and pleural invasion as noted in this case. Other unfavorable factors include pleural effusion at detection, multi-organ involvement, and anemia [3,8].

P-EHE is diagnosed based on histological and immunohistochemical analysis of sampled tissue and can be further evaluated based on molecular characteristics. Histological features include endothelial cells arranged in nests and cords in a myxohyaline stroma. Tumor cells are round with some containing distinct intracytoplasmic vacuolization that results in a signet-ring like appearance. Spindle-shaped tumor cells may also be present [9]. Increased risk of metastasis is associated with marked nuclear atypia, significant mitotic activity and necrosis [3]. As in this patient, immunohistochemical studies are usually positive for CD31, CD34, and Factor VIII-related antigen which is consistent with its vascular origin [10]. Further characterization using molecular studies may be used to confirm the diagnosis; with identification of the WWTR1-CAMTA1 fusion protein helping to differentiate EHE from other vascular tumors [11].

There is currently no consensus on the standard treatment of P-EHE given the rare nature of this condition. Treatment options vary from observation in asymptomatic patients, surgical resection, and chemotherapy in widespread disease. Surgical resection is typically the mainstay for solitary lesions and curative resection has been reported to achieve good outcomes. As described in this case, Radiotherapy (RT) after surgical resection can be used to control localized residual disease; as P-EHE can recur [1,8,12]. Primary RT has been noted to be ineffective in the management of P-EHE due to the slow growth of the tumor and radiobiological features [13]. The use of RT has been most commonly described in EHE of bone following surgical resection. In one case report an RT regimen of 60 Gy in 23 fractions over 43 days for local irradiation of EHE involving the radius in 24 year-old man showed no recurrence or metastasis after serial follow up of the patient for a year [14]. Another report describes employing initial surgical resection for EHE of the mastoid. Subsequent imaging after 18-months of clinical observation revealed evidence of recurrence. Surgical resection was undertaken followed by adjuvant RT. Follow-up revealed no evidence of recurrence or change in clinical status of the patient after eight years [15]. Given the rarity of case reports describing the use of RT for P-EHE, careful clinical follow-up was warranted in the case of our patient and it yielded similar favorable outcomes.

In summary, we describe an elderly patient who was initially treated with surgical resection for a paratracheal mass which was confirmed to be P-EHE; followed three years later with RT due progression of disease involving the pleura. She demonstrated dramatic clinical improvement and no further radiographical progression of her disease, recurrence, or metastases. This case proposes a potentially effective treatment option for P-EHE, with RT as a tool to treat and possibly prevent recurrence of local disease. Further research is required to determine the overall benefits of RT in the treatment of P-EHE, the most effective RT dose for locoregional control, and to characterize the natural history and prognosis of this rare neoplasm.

Learning Points

- EHE is a rare vascular tumor which can arise in different and multiple sites within the body.
- Despite commonly being an incidental finding on imaging, P-EHE may present clinically with symptoms such a consistent, protracted, non-productive cough and shortness of breath.
- P-EHE is typically treated with surgical resection, however, as seen in other variants of EHE; there may be a role for radiotherapy as a potential curative therapeutic intervention.

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