Primary Hemangiopericytoma Arising in Extralobar Pulmonary Sequestration: A Coincidence or Two Rare Disorders?

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Abstract. Background/Aim: Extralobar pulmonary sequestration (EPS) is an unusual congenital defect characterized by the presence of non-functioning lung tissue receiving arterial supply from the systemic arteries. Primary hemangiopericytoma (HPC) is an uncommon potentially malignant tumor of vascular origin that usually involves the soft tissue of the extremities or retroperitoneum, but extremely rarely affects the lung. We present the rare case of a primary pulmonary HPC arising in an EPS. Case Report: A 65-year-old woman, with dyspnea and pleuritic chest pain, was referred for further investigation. Radiological evaluation demonstrated a well-circumscribed mass above the right hemidiaphragm, receiving its arterial supply from the descending thoracic aorta. The patient underwent a right posterolateral thoracotomy and a middle lobectomy. The intraoperative finding was a well-encapsulated solid mass. The histological evaluation described HPC. Results: The patient remains fit and healthy. Conclusion: Pulmonary HPC can arise in EPS. Surgical excision is the treatment of choice.

Extralobar pulmonary sequestration (EPS) is an unusual congenital defect characterized by the presence of non-functioning lung tissue that receives its arterial supply from the systemic arteries, most commonly the thoracic aorta. It is separated from the bronchial tree and has its own pleural covering. Primary hemangiopericytoma (HPC) is an uncommon potentially malignant tumor of vascular origin that usually involves the soft tissue of extremities or retroperitoneum, but extremely rarely affects the lung. We herein present the interesting case of a 65-year-old female patient with primary pulmonary HPC arising in an EPS, thereby implying a possible association which has not been previously reported.

Case Report

A 65-year-old woman, with a 6-month history of dyspnea and pleuritic chest pain, was referred for further investigation of undiagnosed right-sided pleural effusion. She had no relevant medical or family history and had not received any medications recently.
On admission, she had no additional respiratory or systemic symptoms such as cough, fever, fatigue, hemoptysis or weight loss. There were no signs of cyanosis, finger clubbing, peripheral edema, skin lesions or peripheral lymphadenopathy. Chest auscultation revealed diminished breath sounds in the right lower lung zone, while abdominal examination was insignificant. Apart from a mild normocytic anemia (Ht: 30%, Hb: 9.4 g/dl), complete blood count, routine biochemistry blood tests and arterial blood gas examination were within normal limits.

Contrast-enhanced thoracic computed tomography (CT) demonstrated a well-circumscribed mass above the right hemidiaphragm, without evidence of calcification or cavitation, and an accompanying pleural effusion (Figure 1A). The diagnosis of pulmonary sequestration was suspected due to the location of the lesion and the heterogeneous enhancement displayed. Multidetector CT angiography was carried out and confirmed that the mass received its arterial supply from the descending thoracic aorta (Figure 1B-D).

The patient underwent a right posterolateral thoracotomy and a tumorectomy with right middle lobectomy. The intraoperative finding was a sizeable well-encapsulated solid mass, without any presence of normal lung tissue in it, which was successfully detached from the surrounding tissues, and the afferent artery was subsequently ligated (Figure 2). The postoperative period was uneventful. Histological examination of the resected specimen showed infiltration by
regular tumor cells without atypia. The cells presented with round and spindle-shaped nuclei that surrounded vascular channels and had few mitoses per section. Tumor cells were immunohistochemically positive for vimentin, CD34 and CD99, but negative for smooth muscle actin, cytokeratin 14, epithelial membrane antigen and desmin. Staining for factor VIII antigen was positive in lining endothelial cells, but negative in perivascular cells (Figure 3). The diagnosis of primary pulmonary HPC was reliably established based on the above histological findings.

The patient had a significant improvement of her symptoms within the next few months, and remained clinically and radiologically stable, without any signs or symptoms of local tumor recurrence or metastatic disease, 2 years postoperatively.

Discussion

The term HPC was initially suggested by Stout and Murray in 1942 to describe a rare tumor of vascular origin, mainly composed of capillary pericytes (1). Primary pulmonary localization of this type of neoplasm is extremely rare, can occur at any age and may have malignant potential. Benign or malignant differential diagnosis is almost impossible to establish on the basis of clinical examination or plain chest x-rays. Among 36 cases reported in the literature until 1979 (2, 3) and a subsequent case series of 18 patients (4), approximately 40% to 50% of patients with pulmonary HPC were asymptomatic at presentation, despite the impressively large size (≥5 cm) of the tumor in up to two-thirds of cases. Non-specific symptoms such as hemoptysis and chest pain were the commonest, while dyspnea or cough were less frequently met. Large-sized, homogenous, soft-tissue masses with round or slightly lobulated margins and infrequent calcification or no sign of atelectasis were characteristic, though not pathognomonic, radiographic features on x-ray (2, 3). In later studies, CT scan more accurately described calcification in 10% of cases and recognized large central areas of low attenuation in large HPCs, as a radiological indicator of malignancy (5).

Pulmonary sequestration accounts for 0.15-6.4% of all congenital pulmonary anomalies (6). EPS comprises 25% of sequestrations (7). Most cases (approximately 80%) of EPS lie between the lower lobe and the diaphragm, are left-sided and occupy the region of the posterior basal segments (6). The characteristic position of the lesion and recurrent lung infections without significant radiographic resolution in childhood or young adulthood makes intrapulmonary sequestration a possible diagnosis. On the contrary, EPS due to lack of communication with the tracheobronchial tree may remain asymptomatic for many years or less frequently present in the neonatal period with more intense clinical
manifestations such as respiratory failure, cyanosis, infections and/or recurrent pulmonary hemorrhage (8). Contrast-enhanced helical CT scanning with 3-dimensional reconstruction can confirm the suspected diagnosis (9). Conventional or CT arteriography is helpful in differentiating the lesion from other abnormalities of the lung, such as pulmonary arteriovenous fistula, based on the demonstration of systemic arterial blood supply of sequestration (10). Despite the lack of a history of susceptibility to pulmonary infections in our elderly patient, the presence of a well-circumscribed mass located in the lower lung field determined our decision to proceed to CT arteriography and 3-dimensional reconstruction imaging.

The therapy for pulmonary sequestrations is controversial. If asymptomatic, they are often left alone. Even though EPS may show spontaneous regression, the latest study suggests that the volume and diameter of systemic feeding arteries of EPS spontaneously decreased within 4 years without treatment in neonatal patients (11). For those patients with recurrent infections or other symptoms, surgery or embolization or both have been recommended (12). Although the incidence of carcinoma is low, its existence has provided an argument for the need for surgery (13).

The occurrence of benign or malignant neoplasms within pulmonary sequestrations is extremely rare. Nine other cases of primary lung cancer (mainly adenocarcinoma or squamous carcinoma) have already been reported in the English literature. The tumor was found to arise within an intralobar sequestration in all but one case (14). Therefore, the etiology of the carcinomas in the sequestration cases may be multifactorial and may include chronic inflammation and irritation.

There is a single published case of vascular tumor, sclerosing hemangioma, arising within an EPS in a young child (15). A more recent study of a small series of surgically removed intrapulmonary and extrapulmonary sequestrations, revealed hypertensive vascular changes and a possible mechanism of exposure to the elevated pressures of systemic arterial supply was implied (16). Although developmental abnormalities and chronic vascular changes within pulmonary tissue may have a pathogenetic role,
this extremely rare case of HPC development in EPS might be considered a coincidental association.

Radical surgical excision is the treatment of choice for HPC. Intraoperative and postoperative radiotherapy has been proposed as an innovative approach after complete tumor resection (17). The benefits of radiotherapy as palliative treatment for local recurrence or superior vena cava obstruction or chemotherapy for treating patients with metastatic disease or those for whom surgery is not feasible have not yet been clarified (3). Long-term postoperative observation is highly recommended (5), since primary pulmonary HPC may recur locally (pleura, lung, mediastinum) or distally in up to one-third of cases, usually within the first 1-2 years but rarely more than a decade after surgical excision of the tumor (4, 18).

Conflicts of Interest

All the Authors declare that there is no conflict of interest in regard to this study.

Authors’ Contributions

PT designed the study and was the supervisor. PT, CD and DD performed the surgical operation. CFK and CD contributed equally. CFK and CD wrote the article. AAL, NG, AG, IT, ES and AP collected the data. MD performed radiological evaluations. DD and SAP revised the article.

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