CASE REPORT

Human pulmonary dirofilariosis coexisting with intercostal neurilemmoma: A case report and literature review

Chia-Ying Li, Yih-Leong Chang, Yung-Chie Lee

Division of Thoracic Surgery, Department of Surgery, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan

Department of Pathology, National Taiwan University Hospital and National Taiwan University College of Medicine, Taipei, Taiwan

Received 26 November 2009; received in revised form 24 February 2010; accepted 16 May 2010

KEYWORDS

dirofilariosis; neurilemmoma; video-assisted thoracoscopic surgery

Human pulmonary dirofilariosis (HPD) is a rare zoonotic infection caused by *Dirofilaria immitis*. Dogs are the definite hosts and humans are infected occasionally via a vector, generally a mosquito. Most thoracic neurilemmoma arise in the mediastinum and fewer tumors originate peripherally from the intercostal nerves. Most patients with HPD or thoracic neurilemmoma are asymptomatic and these diseases are often discovered incidentally. We present a 53-year-old female who was found to have a pulmonary nodule and a chest wall nodule during a routine health examination. She underwent a video-assisted thoracoscopic surgery (VATS) with partial lung resection and local excision of the chest wall. The pathological examination revealed a coiled, degenerating *Dirofilaria immitis* worm surrounded by granulomatous inflammation with caseous necrosis and a neurilemmoma composed of S-100 protein immunoreactive but smooth muscle actin negative spindle cells. Because these diseases are self-limiting and make further treatment unnecessary, video-assisted thoracoscopic surgery (VATS) is considered preferable and less invasive for definitive diagnosis and management. Copyright © 2012, Elsevier Taiwan LLC & Formosan Medical Association. All rights reserved.

Introduction

Human pulmonary dirofilariosis (HPD) is a rare zoonotic infection caused by an intermediate host, generally a mosquito, which transfers larvae to humans during feeding. The definitive host of the heartworm *Dirofilaria*...
**Dirofilaria immitis** is the dog. Most reported cases of HPD occurred along the Gulf and Atlantic coasts in the US. The frequency and distribution is related to canine dirofilariasis. Overall prevalence of *Dirofilaria immitis* in Taiwan is 57% in stray dogs and 25% in house dogs. However, only two cases of HPD have been reported in Taiwan.

Neurilemmoma is a benign neurogenic tumor that arises from Schwann cells. Most thoracic neurogenic tumors arise in the mediastinum; fewer than 10% of these tumors originate peripherally from the intercostal nerves.

Most patients with HPD or thoracic neurilemmoma are asymptomatic; these diseases are discovered accidentally on routine chest radiography. This report documents the third case of HPD in Taiwan and the first to have received video-assisted thoracoscopic surgery (VATS) for definitive diagnosis and treatment.

**Case report**

A 53-year-old female was admitted for evaluation of a single nodule in the right upper lobe of the lung (Figs. 1 and 2A) and another in the right chest wall (Fig. 2B); both were found incidentally during a routine health examination. She was asymptomatic and had an underlying history of spinal spurs, under rehabilitation. She denied any history of smoking, traveling, or pets. Physical examination, laboratory data and tumor markers were within normal range. Spirometry revealed a forced vital capacity (FVC) of 3.7 L (128% of predicted FVC) and a forced expiratory volume at 1 second (FEV1.0) of 2.61 L (130.2% of predicted FEV1.0).

She underwent VATS right upper lobe wedge resection and pleural tumor excision. A single gray-pink, elastic, oval nodule measuring 0.9 x 0.4 x 0.4 cm in size, over the right upper lobe, and another yellowish-white, elastic firm, oval nodule measuring 1.3 x 1.0 x 1.0 cm in size, over the right chest wall were noted.

Microscopically, the right upper lobe nodule and the adjacent lung parenchyma showed granulomatous inflammation with caseous necrosis. There was a coiled, degenerating *Dirofilaria immitis* worm in the necrotic area, which was highlighted by Grocott's methenamine silver stain (Fig. 3A). The right chest wall nodule revealed a neurilemmoma that was composed of S-100 protein immunoreactive but smooth muscle actin negative spindle cells (Fig. 3B). The patient was discharged on the second postoperative day with no complications and was doing well during follow-up.

**Discussion**

*Dirofilaria immitis* is derived from the Latin words for "evil thread". HPD was first reported in 1887 by De Magalhães in Brazil and the first documented infection resulting in pulmonary infarction was described by Dashiell in 1961.

The dog is the final host where the worm inhabits the right ventricle and is able to grow to approximately 15 to 25 cm in length and 3 to 4 mm in width. Mature adult worms shed several thousand microfilariae per day from the right ventricle into the blood stream. The blood of an infected dog may contain several hundred microfilariae/mm. The microfilariae can be ingested by mosquitoes. They will reach the infectious stage after 10 to 17 days. During the next blood meal, these larvae are deposited on the skin of the next host and then migrate into the subcutaneous tissue. Some species of fleas, ticks, and lice can also act as vectors. These larvae mature for 80 to 120 days in the subcutaneous tissue and then migrate into the circulatory system where they are transported to the right ventricle. They will mature sexually and repeat the cycle later in the right ventricle of the final host.

Cats, foxes, and other mammals can also be hosts.

A human host is a "dead end" host as the larvae cannot develop to a sexually mature form. Their cycle can be arrested in the subcutaneous tissue, resulting in nodular formation. Sometimes, larvae migrate into the human circulation, die in the right ventricle and embolize into the pulmonary artery. Dead larvae will release antigens and cause endarteritis with subsequent pulmonary infarction and granuloma formation.

The frequency and distribution of HPD is related to the prevalence of canine dirofilariasis, the density of vector mosquitoes, and human activities that lead to mosquito bites. A history of dog ownership has little relevance to this disease, as only 25% of patients have ever owned a dog. The distribution of canine infestation is worldwide, especially in stray dogs. In Taiwan, the overall prevalence of canine dirofilariasis is 57% in stray dogs and 25% in house dogs. The prevalence on the main island is 15 times higher than on the offshore islands.

Most documented human cases have been reported from the United States, especially in the Southeast. Only two cases of HPD have been reported in Taiwan, in 1993 and 2003. This report documents the third case in Taiwan and the first to have received VATS for definitive diagnosis and treatment.

Roughly 60% of patients are asymptomatic and usually have a solitary pulmonary nodule noted on radiography. The most common symptom is coughing. Other symptoms,
such as fever, chest pain, hemoptysis, and symptoms resembling the common cold, are rare. Radiographic presentation of HPD is usually a solitary lung nodule with a relatively small diameter (<3 cm in 85% to 95% of patients); 13% of these patients had pleural effusion. Because of the age distribution in the fifth or sixth decade of life and radiographic presentation, it is often difficult to differentiate HPD from primary or metastatic lung cancer.

Systemic eosinophilia has been noted in many parasitic infections, but is rare with HPD (0% to 17%). Other methods of identification, such as skin testing, complement fixation test, examination of bronchial washings, biopsy, or sputum cytology, are of low sensitivity and not helpful. Fine needle aspiration biopsy of the lung and computed tomography-guided aspiration for diagnosis has been performed infrequently. Partial lung resection followed by histopathology is reliable for a definite diagnosis. The characteristic features include a fragment of nonviable *D. immitis* surrounded by eosinophilic pneumonitis and lung infarction. Because the human is a “dead end” host, lesions are always self-limiting and treatment is usually unnecessary. The etiology is the embolization of dead larvae into the distal pulmonary arterial tree. Therefore, most lesions are small and located in the periphery of the lung. VATS with partial lung resection is therefore less invasive and preferable over traditional thoracotomy surgery.

In addition to the HPD, neurilemmoma was found in the lateral chest wall and was thought to originate from the intercostal nerve in this case. Neurilemmoma is the most common neurogenic tumor of the thorax; 90% of thoracic neurogenic tumors originate in the mediastinum, especially the posterior mediastinum. Fewer than 10% arise laterally from intercostal nerves. These tumors are found in persons of any age. They are often asymptomatic and are discovered incidentally on chest radiography or computed tomography. However, the tumors can cause local compression and may lead to symptoms as they become larger in size. Malignant intrathoracic neurogenic tumors are rare: the frequency ranges from 4% to 13%.

Sakai et al reported that magnetic resonance imaging can be used to distinguish neurilemmomas from other thoracic neurogenic tumors. Image-guided, fine-needle cytology, or incisional biopsy can also be used for a definitive diagnosis. However, surgical excision is still considered to be the most acceptable strategy for accurate diagnosis. Local resection is sufficient for small intrathoracic neurogenic tumors; VATS is usually the preferred strategy.

![Figure 2](image1.png)  
(A) Computed tomography revealed a nodule in the right upper lobe (arrow). (B) Another nodule was noted in the right chest wall (arrow).

![Figure 3](image2.png)  
(A) Microscopically, there was a coiled, degenerating *Dirofilaria immitis* worm in the lung parenchyma, which was highlighted by Grocott’s methenamine silver stain. (B) The right chest wall nodule revealed a neurilemmoma that was composed of S-100 protein immunoreactive but smooth muscle actin negative spindle cells.
surgical approach. Larger or more aggressive tumors may require resection of the chest wall.\textsuperscript{11}

In conclusion, HPD is a rare zoonotic infection and is often difficult to differentiate from primary and metastatic lung cancer. The prevalence of canine dirofilariasis is high in Taiwan and all of us have the possibility to deal with this disease. Neurilemmomas are benign and are the most common of intrathoracic neurogenic tumors. For HPD or thoracic neurilemmoma, VATS is considered preferable and less invasive for definite diagnosis and management.

References