

A case of massive fatal hemothorax due to malignant peripheral nerve sheath tumor of the lung

한양대학교 의과대학 내과학교실¹, 한양대학교 구리병원 호흡기내과², 한양대학교 의과대학 병리학교실³

이선민¹, 조민주², 여유미^{1,2}, 박태선^{1,2}, 문지용^{1,2}, 정운용³, 김태형^{1,2}

Introduction: Malignant peripheral nerve sheath tumor (MPNST) is uncommon, biologically aggressive soft tissue sarcoma of neural origin. Treatment for MPNST is challenging with overall poor outcome. We report a case of pulmonary MPNST presented with unilateral spontaneous massive hemothorax.

Case: A 74-year-old woman visited our emergency room with dyspnea of mMRC III for 3 days. She had cured rectal cancer and never smoked. She had no other comorbidity nor recent trauma. At visit, her temperature was 37.2°C; heart rate, 78bpm; blood pressure, 152/81mmHg and respiratory rate, 28bpm. Her chest x-ray showed near total haziness of left lung field and mediastinal shift. Chest CT scan revealed massive hemothorax with possible extravasation from left pulmonary artery and mass like lesion on left lower lobe of the lung. Her hemoglobin level was 8.9 g/dL and ABGA showed Ph 7.28, Pa CO₂ 28.9 mmHg and PaO₂ 63.0 mmHg with FiO₂ 40%. Due to progression of respiratory distress, we performed pigtail catheter insertion and embolization of bleeding focus. After ICU admission and chest tube insertion, hypoxia and respiratory distress continued due to insufficient drainage even after urokinase instillation. Finally, hematoma evacuation via video associated thoracoscopic surgery (VATS) and excisional biopsy for lung mass was done. The biopsy showed high grade sarcoma, compatible with MPNST. The patient expired due to intractable respiratory failure even under mechanical ventilation at Hospital day 28.

Conclusion: Spontaneous massive hemothorax can be caused by various causes. In case of treatment failure even with aggressive bleeding control, we should consider operative procedure for the diagnosis and treatment even with clinical limitations.



Figure 1. Chest x-ray done at the (A) day of admission (A) showed near total atelectasis of the Lt. lung and mediastinal shift to the Right. (B) hospital day 10 after embolization with coil and chest tube insertion. (C) hospital day 12 after hematoma removal and excisional biopsy for lung mass under VATS.

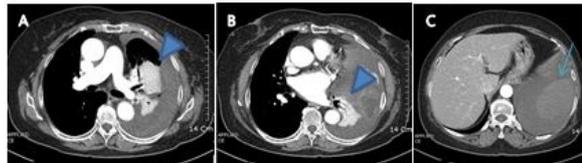


Figure 2. CT scan of chest with contrast showing large left pleural effusion with probable active bleeding of pulmonary arteries (A) upper portion, (arrow head on dye extravasation) (B) middle portion, (arrow head on dye extravasation) (C) lower portion (arrow on Mass-like lesion)

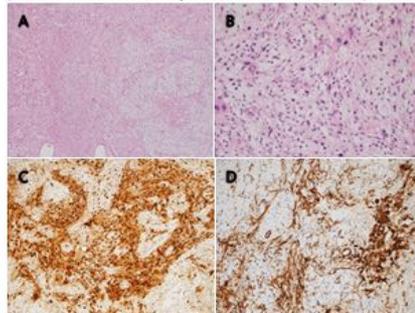


Figure 3. Pathological finding which is diagnosed as high grade sarcoma, consistent with pleomorphic malignant peripheral nerve sheath tumor (A) H&E stain (x100) (B) H&E stain (x400) (C) positive for S-100 protein (x200) (D) positive for smooth muscle actin (x200)