Prognosis of Patients with Idiopathic Pulmonary Fibrosis Admitted to the Intensive Care Unit

Jung-Yien Chien, Jih-Shuin Jerng, Chong-Jen Yu, Pan-Chyr Yang

Background: Idiopathic pulmonary fibrosis (IPF) is a progressive diffuse parenchymal lung disease of unknown etiology. Most IPF patients develop a progressive restrictive lung disease as a result of progressive lung fibrosis, and respiratory failure eventually mandates mechanical ventilation. This study was aimed at studying the clinical features of patients with IPF admitted to the intensive care unit (ICU) due to acute respiratory failure, so as to investigate their mortality and prognostic factors.

Materials and Methods: We retrospectively reviewed the medical records of all patients with IPF admitted to the medical ICU of National Taiwan University Hospital, Taipei, Taiwan, from January 1999 to April 2003.

Results: A total of 13 eligible patients, including 10 men, with a median age of 72 (56-87) years, were identified. Nine patients had been on corticosteroids already at the time of admission. The most common precipitating factor for respiratory failure was pneumonia. Sepsis developed in 11 (85%) patients, while septic shock developed in 6 (46%), and multiple organ failure occurred in 7 (54%). Seven patients (54%) received immunosuppressive therapy in the ICU. The ICU and hospital mortality rates were 62% and 85%, respectively. Univariate analysis showed that the development of septic shock and the increased number of cases of organ failure were associated with significantly higher mortality. Multivariate survival analysis showed that the development of septic shock (hazard ratio=16.69; p=0.04) and the increasing number of organ failure (HR = 3.42 per additional dysfunctional organ system; p=0.04) were independent factors associated with significantly higher mortality.

Conclusions: These findings suggest that IPF patients admitted to the ICU with acute respiratory failure have a poor prognosis in the presence of septic shock or multiple organ failure.
(Thorac Med 2004; 19: 168-177)

Key words: pulmonary fibrosis, respiratory failure, mechanical ventilation

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原發性肺纖維化患者於加護病房之預後探討

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前言：原發性肺纖維化為一不明原因的間質性肺病，罹患此症會導致漸進性肺纖維化和呼吸衰竭。本研究的目的在探討導致患者呼吸衰竭而需要加護病房照顧的原因，其臨床表現及影響預後的因子。

材料與方法：分析台大醫院 1999 年一月至 2003 年四月之間，內科加護病房中罹患原發性肺纖維化患者的臨床資料及預後。

結果：總共有 13 位患者符合原發性肺纖維化的診斷，其中包括 11 位男性，中位數年齡為 72 歲(56-87 歲)。九位患者在住院前已經在使用類固醇類藥物。大部分呼吸衰竭的加重因子為肺炎。七位患者(54%)在加護病房中接受類固醇治療。敗血症發生於 11 位患者(85%)，其中 6 位患者(46%)發生敗血性休克而共有七位患者(54%)產生多器官衰竭。加護病房死亡率為 62% 而住院死亡率增加至 85% 。單變數存活分析顯示住進加護病房前有較長症狀者，若發生敗血症者及較多器官衰竭則會有較高的死亡率。進一步的多變數存活分析顯示發生敗血症(hazard ratio=16.69; p=0.04)及增加器官衰竭數目者(每增加一個器官衰竭 hazard ratio 增加 3.42; p=0.04)是影響預後的獨立因子。

結論：原發性肺纖維化患者因呼吸衰竭而需要加護病房照顧者若發生敗血症及多器官衰竭會有較差的預後。*(胸腔醫學 2004; 19: 168-177)*

關鍵詞：肺纖維化，呼吸衰竭，呼吸器治療

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Heliox-Driven Albuterol Therapy in Severe Persistent Asthma with Acute Exacerbation

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Background: The helium-oxygen mixture (heliox)-powered nebulizer improves the delivery of albuterol to the peripheral lung, but its role in managing acute asthma attacks, however, remains controversial.

Methods: To compare the bronchodilator effect produced by albuterol inhalation delivered by a heliox-powered nebulizer with that by an air-O_2-powered nebulizer in severe asthmatics with acute exacerbation, we conducted a prospective, double-blinded randomized controlled trial in a tertiary university-affiliated medical center in Taiwan. Eighty severe persistent asthmatics (FEV_1 < 60% predicted) with acute exacerbation were randomized to receive 2 doses of albuterol via a nebulizer driven by air-O_2 or heliox (80:20). All patients received intravenous methyl-prednisolone. Arterial blood gas (ABG), hemodynamics, spirometric data, and dyspnea scores were collected before and after albuterol treatments.

Results: Patients receiving heliox-driven albuterol had a higher peak expiratory flow rate (PEFR) (heliox vs. air-O_2 = 44.9 ± 6.1% vs. 38.4 ± 7.8% after the 2nd treatment, p< 0.01), and a lower dyspnea score (heliox vs. air-O_2 = 2.4 ± 1.1 vs. 4.0 ± 1.1 after the 2nd treatment, p <0.01).

Conclusion: We conclude that albuterol delivered by a heliox-driven nebulizer may be a useful adjunct therapy in severe persistent asthmatics with acute exacerbation. ***(Thorac Med 2004; 19: 178-186)***

Key words: Helium, bronchial asthma, peak expiratory flow rate, dyspnea score, forced expiratory volume in 1st second.

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氦氧混合氧驅動下對重度氣喘急性發作病患使用支氣管擴張劑化霧治療時的影響

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過去文獻顯示氦氧混合氣可改善乙型支氣管擴張劑 albuterol 到肺部的吸收量，但它對臨床氣喘發作者的療效，仍無結論。本文即針對氣喘重度發作病患，分別以氦氧混合氣（實驗組）和氧氣（對照組）驅動 albuterol 化霧治療後，比較其對支氣管擴張的療效影響的差異。

方法：本文將八十位急性重度氣喘發作病患（FEV_1 < 60% 預估值）分成兩組。分別接受兩次以氦氧混合氣（80：20）或氧氣驅動 albuterol 做化霧治療。所有病患均接受靜脈注射類固醇，在治療前後均收集病患動脈血的氣體分析、血流動力學、肺功能以及急促參數，來比較分別以氦氧混合氣和氧氣驅動 albuterol 對支氣管擴張療效的差異。

結果：以氦氧混合氣驅動 albuterol 化霧治療下的氣喘病患組，呈現統計學上明顯較佳的最大吐氣流量比率（實驗組：對照組＝ 44.9 ± 6.1%：38.4 ± 7.8% ; p <0.01）以及較佳的急促指數（實驗組：對照組＝ 2.4 ± 1.1：4.0 ± 1.1; p<0.01）。

結論：本文研究顯示，對於重度氣喘發作病患，施予氦氧混合氣驅動支氣管擴張劑之化霧治療可提供一有效的輔助工具。***(胸腔醫學 2004; 19: 178-186)***

關鍵詞：氦氣，氣喘，最大吐氣流量，急促指數，第一秒最大吐氣量

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Mediastinal Cystic Lymphangioma — A Case Report and Literature Review

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Cystic lymphangiomas are extremely rare mediastinal tumors in adult patients. They are usually located in the neck (75%) or axillary region (20%), with only 1% localized in the mediastinum. When present in the mediastinum, they can be located in all mediastinal compartments, with a predilection for the anterior mediastinum. The tumor is usually noticed when compression symptoms occur, or when it is accidentally found on the chest X-ray. Complete surgical resection is indicated for this tumor. We present a case and discuss the occurrence, pathogenesis, and management of the postoperative chylothorax of a cystic lymphangioma.
(Thorac Med 2004; 19: 187-193)

Key words: cystic lymphangioma, chylothorax

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Mediastinal Cystic Lymphangioma

縱膈腔囊性淋巴管瘤

張仲元　*馮瑤　**蘇正川　鄭高珍

淋巴管瘤發生在成人縱膈腔是非常罕見。其發生部位，在頸部佔 75%，腋窩部佔 20%，而縱膈腔祇有 1%。淋巴管瘤可以在縱膈腔任何部位發生，但好發於前縱膈腔。縱隔淋巴管瘤主要是在胸部 X 光檢查時意外發現，或發生壓迫症狀時才被發現。處理方法是以完全手術清除為主。這裡提供一個病例，並討論其發生率，病理及手術後乳糜胸的處理。*(胸腔醫學 2004; 19: 187-193)*

關鍵詞：囊性淋巴管瘤，乳糜胸

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Bronchoscopic Findings of Post-intubation Tracheal Rupture in Critically Ill Patients: Report of Four Cases and Review of the Literature

Che Kim Tan, Jih-Shuin Jerng, Kuan-Yu Chen, Chong-Jen Yu, Pan-Chyr Yang

Tracheal rupture is a rare but potentially life-threatening complication of endotracheal intubation. We present four cases of post-intubation tracheal rupture with various bronchoscopic findings. One of the ruptures was caused by a direct stylet injury. All of the patients were elderly women (82 ± 8 years) with critical medical conditions (APACHE II score 35 ± 5) who underwent emergency intubation due to respiratory failure. None of them had pneumothorax. Subcutaneous emphysema was the earliest sign, noted at 3.1 ± 2.3 hours after intubation. All patients experienced a delayed diagnosis (5.3 ± 1.9 days), and lengthy ICU (24 ± 22 days, median 15 days) and hospital (46 ± 34 days, median 38 days) stays. The mortality rate was high (50%). We offer a suggestion on how to avoid this complication, and recommend a detailed physical examination for subcutaneous emphysema for every intubated patient in order to obtain an early diagnosis. Post-intubation subcutaneous emphysema should mandate a chest computed tomography and bronchoscopic evaluation for this potentially life-threatening condition. ***(Thorac Med 2004; 19: 194-201)***

Key words: endobronchial intubation, tracheal rupture, bronchoscopy, subcutaneous emphysema

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Bronchoscopic Findings of Post-intubation Tracheal Rupture

重症病患之氣管插管後併發氣管破裂的支氣管鏡檢表現：四例病例報告及文獻回顧

陳志金　鄭之勛　陳冠宇　余忠仁　楊泮池

氣管破裂乃氣管插管罕見但可能致命的併發症。我們在此報告四例氣管插管後併發氣管破裂的支氣管鏡檢下的不同表現。其中一例之氣管破裂直接由氣管內管通條所致。四位病患皆為高齡女性(82 ± 9 歲)且病況嚴重(APACHE II 分數 35 ± 5)。四位皆因呼吸衰竭而接受緊急氣管插管。四位皆無併發氣胸。皮下氣腫是最早的表徵，在插管後 3.1 ± 2.3 小時被發現。我們發現有明顯的診斷延誤(5.3 ± 2.0 天)，超長的加護病房住院天數(24 ± 22 ，中數， 15 天)及總住院天數(46 ± 34 ，中數， 38 天)。死亡率則高達 50%。因此，我們對於插管前的步驟作一些建議以減少此一併發症的發生，並建議針對氣管插管後病人作完整的理學檢查以便及早發現皮下氣腫以利早期診斷氣管破裂。當氣管插管後發生皮下氣腫，應進一步安排胸腔電腦斷層掃描及支氣管鏡檢查。*(胸腔醫學 2004; 19: 194-201)*

關鍵詞：氣管插管，氣管破裂，支氣管鏡檢查，皮下氣腫

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Primary Mediastinal Yolk Sac Tumor — A Report of Two Cases and Literature Review

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Yolk sac tumors, also called endodermal tumors, are a kind of germ cell tumor, mostly found in the gonad. Extragonadol germ cell tumors are rare, and are usually found in the mediastinum. Mediastinal tumors are more prevalent in young males. Clinical presentations including chest pain, dyspnea, cough and body weight loss are noted, though asymptomatic cases with a huge anterior mediastinal mass, disclosed by chest radiography, are also seen. Due to the small sample size available for pathological study, yolk sac tumors are often misdiagnosed and consequently mismanaged.

We came across 2 cases of primary anterior mediastinal tumors referred from a local hospital, where the initial diagnoses by fine needle aspiration biopsy were undifferentiated adenocarcinomal. However, at our hospital the markedly elevated serum alfa-fetoprotein (AFP) and normal β-human chorionic gonadotropin (β-hCG) of these two young males led to a suspicion of yolk sac tumors before surgical intervention. After surgical resection of the tumor, the specific pathologic characteristics and immunohistochemical staining confirmed the yolk sac tumor diagnosis. We present these 2 cases and review the associated literature. ***(Thorac Med 2004; 19: 202-208)***

Key words: yolk sac tumor, germ cell tumor, mediastinal mass

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前縱膈腔原發卵黃瘤之二病歷報告及相關文獻回顧

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卵黃囊瘤是屬於 Germ cell tumor 的惡性非畸胎瘤(nonteratomatous)一種，絕大部份原發於生殖腺，極少數會有原發於生殖腺外，其中以縱膈腔內為常見，病患年齡分佈以年輕男性居多，病患常以胸痛，呼吸困難，咳嗽及體重減輕來表現，有些是毫無徵狀，是在常規身體檢查意外發現。其胸部 X 光發現常是典型之縱膈腔腫瘤來表現，在胸部電腦斷層上是呈現不均質腫瘤，在打顯影劑後可能會有不均質或邊緣顯影，至於病理診斷方面，若僅以病理切片之表現尤其是細針抽吸切片，可能會誤判為分化不良型腺癌，須配合血清胎兒蛋白上升及切片之免疫染色，有時須藉助外科手術以取得適當檢體也同時儘量切除腫瘤，以利於以 platinum 為基礎之化學治療的進行。

最近我們連續遇到兩名病人其在外院診斷為分化不良型腺癌而在本院最終診斷是前縱膈腔卵黃囊瘤，藉這兩個病例描述其臨床表現，影像學發現，診斷及治療經過並回顧歷年來與此種病歷相關的文獻報告。
(胸腔醫學 2004; 19: 202-208)

關鍵詞：卵黃囊瘤，分化不良型腺癌，縱膈腔腫瘤

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Pseudomembranous Tracheobronchitis Caused by *Aspergillus* in an Immunocompromised Patient — A Case Report

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Invasive aspergillosis is a leading cause of death in immunocompromised patients. We herein report a case of pseudomembranous tracheobronchitis caused by Aspergillus in an immunocompromised patient. A 45-year-old woman with diabetes mellitus presented with chest pain and cough. She was found to be in diabetic ketoacidosis and was treated for that and possible respiratory sepsis. She subsequently developed airway obstruction and respiratory failure. Bronchoscopic examination revealed pseudomembranous tracheobronchitis with granulation tissue obstructing several bronchi. The pseudomembrane consisted histologically of septate hyphae branching at 45°, consistent with *Aspergillus* species. Despite mechanical ventilation and antifungal therapy, the patient succumbed to progressive obstructive respiratory failure. Aspergillus tracheobronchitis should be considered in immunocompromised patients presenting with cough, chest pain, fever, dyspnea and upper airway obstruction. Early bronchoscopy and histologic examination should be performed and all clinical isolate of Aspergillus spp should be identified to species level because early definitive treatment may be life - saving. ***(Thorac Med 2004; 19: 209-214)***

Key words: diabetes mellitus, aspergillus, pseudomembranous tracheobronchitis, ketoacidosis, obstructive respiratory failure

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麵菌病會造成免疫力失調病人產生僞膜性氣管支氣管炎—病例報告

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侵略性的麵菌病 (Invasive aspergillosis) 是造成免疫力失調病人死亡的主要原因。我們提出一份報告有關患有麵菌病僞膜性氣管支氣管炎 (Aspergillus Pseudomembranous tracheobronchitis) 之免疫力失調病人的臨床病例，一位中年45歲患有糖尿病 (Diabetic mellitus) 的婦人，她初期呈現胸痛、咳嗽及呼吸困難的情況，我們將她視為酮酸症 (Ketoacidosis) 及敗毒病作治療。之後她又產生氣道呼吸障礙的情形，在進行支氣管透視鏡檢查後顯示因粗糙組織造成僞膜性氣管支氣管炎 (Pseudomembranous tracheobronchisis) 有支氣管障礙的情形發生。僞膜性氣管支氣管炎 (Pseudomembranous tracheobronchisis) 是由有隔膜分支 hypae 的組織構成，此是與麵菌病 (Aspergillosis) 的種類相同的。即使進行人工換氣或抗菌治療，她還是死於因急速的呼吸障礙功能失調 (Obstructive respiratory failure)。因此結論是當一個免疫力失調病人產生咳嗽、胸痛、發燒及呼吸困難的情況時，我們應該要立刻察覺麵菌病僞膜性氣管支氣管炎 (Aspergillus pseudomembranous tracheobronchisis) 的存在。早期的支氣管透視鏡檢查及組織檢驗，能早期接受治療延長存活率。*(胸腔醫學 2004; 19: 209-214)*

關鍵詞：麵菌病 (Aspergillosis)，僞膜性氣管支氣管炎 (Pseudomembranous tracheobronchisis)，糖尿病 (Diabetic mellitus)，酮酸症 (Ketoacidosis)，呼吸障礙功能失調 (Obstructive respiratory failure)

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Primary Malignant Fibrous Histiocytoma of the Lung: A Case Report

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Primary malignant fibrous histiocytoma (MFH) arising in the lung is a rare clinical entity. We report a 34-year-old female who presented with a two-month history of dry cough. There was no fever, chest pain, shortness of breath, or body weight loss. Earlier chest roentgenograms revealed a cavity nodule in the left upper lobe, and no interval change in the size two months later. The chest computed tomography (CT) revealed a nodular lesion about 2.5 cm in the left upper lobe with partial cavity formation. The patient underwent CT-guided biopsy and the pathology reported a neoplasm. The possibility of malignancy was considered due to hypercellularity and cellular atypia. A left upper lobe lobectomy was performed and the final pathological diagnosis was malignant fibrous histiocytoma without lymph node metastasis. MFH is the most common soft tissue sarcoma in adults, usually arising in the extremities or trunk. The chest is a rare primary location for this tumor. ***(Thorac Med 2004; 19: 215-219)***

Key words: malignant fibrous histiocytoma (MFH), soft tissue sarcoma

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Primary Malignant Fibrous Histiocytoma of the Lung

原發性肺臟惡性纖維組織細胞瘤：病例報告

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原發性肺臟惡性纖維組織細胞瘤臨床上非常罕見，本篇報告壹位三十四歲女性主訴為?咳達兩個之久，沒有合併發燒，胸痛，呼吸急促或體重減輕。 CXR 顯示一開洞結節位於左上肺葉，胸部電腦斷層顯示此結節約2.5 公分，有部分開洞形成。電腦斷層下的切片結果疑似惡性腫瘤。我們施行左上肺葉切除手術，病理報告顯示為肺臟惡性纖維組織細胞瘤。惡性纖維組織細胞瘤是最常見的成人軟組織肉瘤，通常位於四肢或軀幹。此病例沒有在其它地方發現軟組織肉瘤存在。位於肺臟的原發性惡性纖維組織細胞瘤相當少見，我們報告這一罕見病例並作文獻回顧與討論。*(胸腔醫學 2004; 19: 215-219)*

關鍵詞：惡性纖維組織細胞瘤，軟組織肉瘤

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Mycotic Pseudoaneurysm of the Radial Artery Occurring after Percutaneous Arterial Cannulation — A Case Report

Chien-Sheng Huang, Han-Shui Hsu

The indwelling percutaneous radial artery catheter is used worldwide especially during major operations or with critical patients in the intensive care units. Complications that occur following radial artery puncture for continuous blood pressure monitoring and frequent arterial blood gas determinations include vascular insufficiency, bleeding, and infection. Most of these complications can be managed conservatively, without deficit. We report a rare complication, a mycotic pseudoaneurysm, occurring after arterial catheterization, which necessitated radial arterial ligation and skin grafting. ***(Thorac Med 2004; 19: 220-223)***

Key words: mycotic pseudoaneurysm, complication, arterial cannulation

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Mycotic Pseudoaneurysm of the Radial Artery

施打周邊動脈導管併發罕見病菌性僞動脈瘤—病例報告

黃建勝　許瀚水

周邊動脈導管插入是呼吸加護病房最常用來血壓偵測及動脈血氣體分析的一種侵入性處置。常見的併發症包括血管的傷害、出血、以及感染。感染性僞動脈瘤是屬於罕見的感染性血管損傷，通常與金黃色葡萄球菌感染有關。其處理要點在病人有危險因子及可疑症狀時須高度的懷疑，同時及早採取適當之治療措施。在此我們提出一例因外科手術而住入呼吸加護病房，住院期間因周邊動脈導管感染而併發病菌性僞動脈瘤的例子。*(胸腔醫學 2004; 19: 220-223)*

關鍵詞：病菌性僞動脈瘤，併發症，動脈導管

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Intrathoracic Neurilemmoma of the Vagus Nerve: A Report of Two Cases

Joseph Ching Li, Hong-Chung Wang, Jau-Yeong Lu, Kuo-An Chu, Jyh-Seng Wang

Although intrathoracic neurogenic tumors are not uncommon, particularly in the posterior mediastinum, very few reports concerning the intrathoracic neurilemmoma arising from the vagus nerve have appeared in the past decades. We report on two cases of intrathoracic neurogenic tumors of the vagus nerve. The chest roentgenograms of these two patients demonstrated a mass in the superior mediastinum. The chest computed tomography scan showed well-defined masses in the right superior mediastinum, with homogeneously low attenuation. The post-contrast image demonstrated a relatively heterogeneous enhancement with smooth capsulation. Both patients received a complete excision, and the pathologic reports of neurilemmoma were identical. The tumors exerted a compressive effect on the surrounding mediastinal structures, resulting in Horner's syndrome and chest pain in the first patient, and chronic non-productive cough in the second. Horner's syndrome is not only associated with postoperative complications, but is also a sign of a recurrent neurogenic tumor. ***(Thorac Med 2004; 19: 224-229)***

Key words: neurilemmoma, neurogenic tumor, Horner's syndrome, vagus nerve

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Vagus Nerve and Neurilemmoma

胸廓內迷走神經之神經鞘瘤：二病例報告

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胸腔內神經瘤並非少見，絕大部份位於後縱膈腔，而胸廓內迷走神經腫瘤則相當稀少，現在我們提出二病例，病患胸部X-光片呈現一均勻軟組織影像於右上前縱膈腔，電腦斷層影像顯示邊緣清楚，密度不均勻且對比劑後顯影不明顯的腫塊，兩者經接受手術切除後證實為右側迷走神經之神經鞘瘤。該類腫瘤可長大壓迫上縱隔膜的組織而造成症狀，其一病患輕微胸痛合併Horner's症候群多年。另外一病患則有慢性乾咳。Horner's症候群不但可能是頸部或上縱膈腔腫瘤手術後的併發症，亦可能是一個原發式腫瘤復發的重要表徵。*(胸腔醫學 2004; 19: 224-229)*

關鍵詞：迷走神經、Horner's症候群、神經鞘瘤、神經源性腫瘤

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Intrapulmonary Arteriovenous Malformation in a Patient with Type I Gaucher's Disease

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Pulmonary involvement is rare in type I Gaucher's disease (GD). Herein, we report a 34-year-old woman who developed pulmonary arteriovenous malformation (AVM) ten years after Gaucher's disease had been diagnosed. Splenomegaly, bone pain, and bruising easily were present in her childhood. Dyspnea developed 10 years after diagnosis of Gaucher's disease. The chest radiograph revealed a tortuous tubular structure in the right lower lung, then echocardiography with a bubble test and cardiac catheterization confirmed the diagnosis. She underwent coil embolization by cardiac catheterization, and her dyspnea improved. Possible mechanisms of intrapulmonary AVM in Gaucher's disease are discussed. ***(Thorac Med 2004; 19: 230-235)***

Key words: intrapulmonary AVM, Gaucher's disease, coil embolization

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Intrapulmonary AVM in Gaucher's Disease

第一型高雪氏症合併肺內動靜脈畸型

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肺部的侵犯在第一型高雪氏症 (Type I Gaucher's disease) 患者身上是罕見的。我們在此提出一個三十四歲的女性病例，她在十年前被診斷為高雪氏症，十年後發生肺部的動靜脈畸型 (pulmonary AVM)。在孩童時期，此病患有脾臟腫大，骨頭疼痛，以及皮膚容易瘀血的現象。在高雪氏症被診斷的十年後，此病患出現呼吸喘的症狀。胸部影像學檢查，發現在其右下肺葉出現一個扭曲的管狀結構之影像。經心臟超音波顯影泡沫試驗 (echocardiography with bubble test) 以及心導管 (cardiac catheterization) 確定診斷為肺動靜脈畸型。此病患接受由心導管所施行的線圈栓塞術 (coil embolization) 後，呼吸喘的症狀大大改善。我們在此討論肺動靜脈畸型在高雪氏症病患中發生的可能基轉。*(胸腔醫學 2004; 19: 230-235)*

關鍵詞：肺內動靜脈畸型，高雪氏症，線圈栓塞術

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