18.14.9 Lipoid (lipid) pneumonia 4263 S.J. Bourke

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18.14.9 Lipoid (lipid) pneumonia 4263 Other manifestations Exudative pleural effusions can occur from amyloid deposits directly involving the pleura. Transudative pleural effusions and pulmonary oedema are often complications of amyloid cardiomyopathy or neph- rotic syndrome. Respiratory muscle weakness has been reported due to infiltration of the diaphragm and skeletal muscles by amyloid. Lymph node enlargement may be seen on CT images. Macroglossia may cause or aggravate obstructive sleep apnoea. The pulmonary vas- culature often contains deposits of amyloid at post-mortem examin- ation. This is usually of no clinical consequence but has been reported to cause pulmonary hypertension and may be associated with an in- creased risk of bleeding when amyloid tissue is biopsied. Management and prognosis Local deposits of amyloid in the larynx, trachea, or bronchi may require treatment by endoscopic interventions, with mechanical debulking by forceps resection or laser therapy. This is effective in re- lieving airway obstruction, but there is a risk of provoking bleeding and recurrence is common. Stenting may also be used to maintain airway patency. Radiotherapy has also been deployed successfully, and in certain circumstances may be a better option with less risk of bleeding or recurrence. Amyloid deposits in the lung parenchyma often do not give rise to symptoms and may not require treatment, but they simulate bronchial carcinoma and may therefore be resected. Systemic AL amyloidosis may be treated by chemotherapy, using agents such as melphalan to reduce the production of immunoglobulin light chains. Diffuse alveolar amyloidosis has a poor prognosis, particularly be- cause it is associated with cardiac and renal amyloidosis. Lung trans- plantation has very rarely been performed for pulmonary amyloidosis. FURTHER READING Borie R, et al. (2012). Tracheobronchial amyloidosis: evidence for local B-cell clonal expansion. Eur Resp J, 39, 1042–5. Gillmore JD, Hawkins PN (1999). Amyloidosis and the respiratory tract. Thorax, 54, 444–51. Jaccard A, et al. (2007). High-dose melphalan versus melphalan plus dexamethasone for AL amyloidosis. N Engl J Med, 357, 1083–93. Lachmann HJ, et al. (2007). Natural history and outcome in systemic AA amyloidosis. N Engl J Med, 356, 2361–71. McLaughlin AM, et al. (2006). Amyloidosis in cystic fibrosis: a case series. J Cyst Fibrosis, 5, 59–61. Milani P, et al. (2017). The lung in amyloidosis. Eur Respir Rev, 26, 170046. doi.org/10.1183/1600617.0046-2017. Neben-Wittich MA, Foote RL, Kalra S (2007). External beam radiation therapy for tracheobronchial amyloidosis. Chest, 132, 262–7.

Pinney JH, Lachmann HJ (2011). Amyloidosis and the lung. Eur Respir Mon, 54, 152–70. Santiago RM, Scharnhorst D, Ratkin G, Crouch EC (1987). Respiratory muscle weakness and ventilatory failure in AL amyloidosis with muscular pseudohypertrophy. Am J Med, 83, 175–8. Ware LB, et al. (1998). Lung transplantation for pulmonary amyloid- osis: a case report. J Heart Lung Transplant, 17, 1129–32. 18.14.9 Lipoid (lipid) pneumonia S. J. Bourke ESSENTIALS Exogenous lipoid pneumonia occurs when animal, vegetable, or mineral oils are aspirated or inhaled into the lungs, provoking a for- eign body reaction with chronic inflammation. Typical symptoms are cough and breathlessness. The chest radiograph and CT may show interstitial thickening, with areas of consolidation that may coalesce into a mass (paraffinoma) which simulates carcinoma. Bronchoalveolar lavage and biopsy show lipid-laden macrophages. In endogenous lipoid pneumonia the lipids are derived from surfactant and cholesterol released from decaying cells distal to bronchial obstruction. Introduction Lipoid pneumonia is an unusual form of lung disease resulting from the accumulation of lipids in the alveoli, where they provoke a for- eign body reaction with associated inflammation and sometimes local fibrosis. The lipids may be endogenous or exogenous in origin, and the clinical mechanisms and circumstances differ accordingly. Endogenous lipoid pneumonia is usually part of another lung dis- ease, notably bronchial obstruction from carcinoma or bronchio- litis, where cholesterol and lipid–rich surfactant accumulate in the alveolar macrophages. Exogenous lipoid pneumonia is caused by the aspiration or inhalation of animal, vegetable, or mineral oil, or hydrocarbons such as petroleum-based substances (Box 18.14.9.1). When ex- ogenous mineral or vegetable lipids are deposited in the lung, they Fig. 18.14.8.2 Alveolar–interstitial type amyloidosis of the lung. Staining with Congo red stain under polarized light (high magnification) demonstrates the characteristic dichroic birefringence. By courtesy of Dr T. Ashcroft.

 are usually relatively inert but difficult to remove. Lung lipases have little effect, and macrophages are slow to transport the free or emulsified material into the lymphatics, such that they are re- tained in the lung for a long time. The result is often a chronic low-grade inflammatory response that may lead to local fibrosis. Animal lipids are more readily degraded by lung lipases, releasing irritating fatty acids and causing a brisk and more widespread pneumonitis, particularly if lipid material is inhaled in large quantities. Exogenous lipoid pneumonia Aetiology Acute lipoid pneumonia Acute lipoid pneumonia is the result of an episode of massive ex- posure to petroleum-based products. This usually occurs as a result of an accident or specific circumstances, such as ‘fire eater’s lung’ or ‘petrol-siphoner’s lung’. In fire eating the performer blows out a mouthful of a flammable petroleum-based liquid against a burning stick, creating an aerosol that burns around the stick. In siphoning petrol, fluid may be sucked into the mouth and aspirated during in- gestion or when vomiting. Shipwrecked sailors have suffered lipoid pneumonia from aspirating floating oil in the sea. A high level of lipid deposition in the lung can produce severe acute respiratory failure, as well as systemic effects involving other organs. Chronic exogenous lipoid pneumonia Chronic exogenous lipoid pneumonia is an indolent disease re- sulting from repeated aspiration or inhalation of lipids into the lung. Diagnosis is based upon identifying a history of exposure to exogenous lipid, and this has been described in a diverse range of settings and circumstances. In infants and children exogenous lipoid pneumonia has occurred as the result of feeding debilitated mal- nourished children with milk, ghee (clarified butter), or liquids with a high lipid content. Administration of cod liver oil to reluctant chil- dren has also resulted in deposition of lipid in the lungs. Sometimes repeated use of petroleum-based nasal jelly is the source of the ex- ogenous lipid. In parts of India there was a common practice of cleaning the mouth, throat, and nose of infants with oil, and this resulted in a high incidence of lipoid

pneumonia. In older people lipoid pneumonia has most often occurred from the regular use of li- quid paraffin as a laxative, taken each night for chronic constipation. Nasogastric feeding can also result in repeated deposition of lipid in the lungs. Often there are associated problems which predispose to aspiration, such as an impaired cough reflex, gastro-oesophageal reflux, or neurological disease, causing difficulties in swallowing or impaired consciousness. In the occupational setting lipid inhalation may occur in fire- fighters exposed to oil mists and burning fats, and in factory workers inhaling metal working fluid contaminated with lubri- cant oils. Local customs and habits may cause lipid pneumonia in particular circumstances, such as the blackfat tobacco smokers of Guyana. Blackfat is a tobacco leaf to which mineral oil and Vaseline are added for flavouring, leading to recurrent inhalation of lipids. Clinical features The clinical features depend on the irritant properties of the lipid material and the dose retained in the lung, and whether any add- itional materials have been aspirated into the lungs. Chronic low-grade aspiration of lipid often produces no imme- diate symptoms and the affected subject may present by chance with an abnormal chest radiograph. However, in about 50% of cases there is a chronic pneumonic illness with productive cough, low-grade fever, and occasionally haemoptysis. Repeated aspiration may lead to fibrotic shrinkage of the affected segments, usually in the lower lobes or the middle lobe. Sometimes the radiographic appearances may closely simulate bronchial carcinoma and surgical resection may be undertaken, revealing a characteristic granulomatous mass (paraffinoma). When more substantial quantities are aspirated the radiographic abnormalities are more diffuse, and when the lipid material is more reactive an acute pneumonic illness occurs. CT may allow the iden- tification of lipid material by its low density (similar to body fat, – 150 to –80 Hounsfield units, compared with + 50 to + 150 units for solid tumours) and also show patchy areas of ground-glass attenu- ation and interstitial thickening, thereby producing a ‘crazy paving’ pattern. Diagnosis is crucially based on identifying exposure to ex- ogenous lipid, and this often requires a detailed history, con- centrating on particular risk factors. Identifying lipid material and lipid-laden macrophages in bronchoalveolar lavage fluid or sputum is helpful, but this must be interpreted in the context of the full clinical circumstances and the presence of any diseases asso- ciated with endogenous lipoid pneumonia. Transbronchial biopsy or surgical lung biopsy may be needed, and typically shows lipid- laden macrophages, multinucleated giant cells, and interstitial fi- brosis (Fig. 18.14.9.1). Box 18.14.9.1 Lipoid pneumonia Exogenous lipoid pneumonia Aspiration • Liquid paraffin laxatives • Paraffin-based nasal jelly • Cod liver oil • Milk feeds • Ghee feeds • Nasogastric feeding • Cosmetic oils • Petrol-siphoner’s lung Inhalation • Metal working fluid • Oil mists • Blackfat tobacco smoking • Fire fighters • Fire eater’s lung Endogenous lipoid pneumonia • Bronchial obstruction • Bronchiolitis • Niemann–Pick disease • Fat embolism

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