Pulmonary nodules due to reactive systemic amyloidosis (AA) in Crohn’s disease

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Abstract

Multiple nodules of AA (reactive systemic) amyloid were identified at necropsy in the lungs of a patient with Crohn’s disease. No other organs were involved. Nodular pulmonary amyloidosis is usually caused by deposition of AL (primary) amyloid.

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Nodular pulmonary amyloidosis is an infrequent manifestation of amyloid disease. In this report multiple nodules of amyloid were identified at necropsy within the lungs of a patient who had active Crohn’s disease but no amyloid deposits at other sites.

Case report

A 75 year old woman presented with a two month history of diarrhoea and two stone weight loss. She had features of congestive cardiac failure and her chest radiographs showed diffuse basal shadowing only, obscuring any further disease. Three days after hospital admission the patient sustained a fatal cardiac arrest.

There was no history of drug ingestion and no significant past medical history, but she was a heavy smoker.

At necropsy no lesions of the tongue or skin were seen. Recent myocardial infarction was confirmed and each pleural cavity contained 500 ml of clear effusion fluid. The lungs were oedematous, together weighing 1480 g. Throughout both lungs, but predominantly in the bases, there were 12 well circumscribed nodules of firm, brown, waxy material up to 3 cm in diameter. There was a terminal ileitis and patchy colitis with a cobblestone mucosal pattern and areas of fibrous stenosis.

Histological examination showed that the pulmonary nodules were composed of amyloid with scattered foci of calcification and ossification. They stained pink with Congo red eliciting apple-green dichroism in polarised light (figure). Staining was abolished by pretreatment of the sections with potassium permanganate. Immunohistochemical examination with the avidin-biotin complex method showed positive staining for amyloid A component (Dako, UK). The ileum and colon showed features of active Crohn’s disease.

Congo red staining of all major organs (including myocardium, kidney, bladder, liver, spleen, small and large intestine, adrenal, thyroid, and pancreas) revealed no evidence of amyloid deposition.

Discussion

Nodular pulmonary amyloidosis is an uncommon but recognised manifestation of amyloid
Crohn's disease is infrequently complicated by amyloidosis but usually follows the pattern of reactive systemic amyloidosis. In the absence of an alternative explanation it is suggested that the unusual amyloid deposits in this patient occurred as a consequence of Crohn's disease.

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BOOK NOTICE


This is the second edition of a small handbook intended for medical and nursing staff, both at student and practising levels. It is stated in the preface that the subject matter is tied by the common thread of applied respiratory physiology, and indeed this book does provide a practical and didactic guide to care of the respiratory patient with a strong emphasis on their physiological defects and their management. This is a welcome deviation from the more common diagnosis based approach to respiratory disease and provides an introduction, as the book's title suggests, to the practices required in respiratory intensive care. Although there is much useful information within this manual, it fails to be consistent in its assumption of basic knowledge; in some places so little knowledge is assumed that the descriptions become extremely laboured—for instance, the description of partial pressures in the opening chapter—whereas in other parts limited information is given and presumably the reader is expected to have the knowledge. Furthermore, the balance of information seems to be inadequate in some places—for instance, in the third chapter on water and solute balance there is very limited description of capillary water movement and an extremely detailed description of cell water movement. Similarly, the chapter on diagnosis of respiratory disease provides an excellent description of the physiology behind respiratory symptoms and signs, but gives very little detail to the physiology behind respiratory function tests radiography.

The book opens with chapters on anatomy and physiology, the physiology of water and solute balance, acid base physiology, and a physiological approach to respiratory diseases tying together the basics very well. The chapter on respiratory failure again covers the physiological aspects very well, but the reader is left with very little information on how to manage respiratory failure. The management of the airway is covered in two excellent chapters dealing with methods of keeping the airway open and methods of improving airway clearance. Therapeutics are discussed with a single chapter on drugs, although drug interactions are poorly covered. The use of paralysing agents in intensive care is covered, but sedatives are not mentioned. Although most drugs are covered in this single chapter, aerosol therapy is given a very full and detailed chapter of its own. Mechanical ventilation is discussed in a chapter which opens with ways of avoiding mechanical ventilation and gives a limited description of the various modes of ventilation. Useful in a respiratory manual is a chapter dealing with the disposition of oxygen once it has crossed the lungs; related to this there is an excellent chapter on the "whys and wherefores" of monitoring. There are some glaring omissions. Infection control is covered in a full chapter dealing with transmission of HIV disease but the common respiratory problems of HIV disease are not mentioned. The reader who expects this book to be a respiratory intensive care manual will be disappointed. Those who expect what the title literally promises should be reasonably satisfied. This point is emphasised with an excellent chapter on home respiratory care, covering home oxygen therapy and home ventilation with useful information on patient selection and support.

In conclusion, this book has a place on the bookshelves of medical students and junior doctors. It is an additional source of information that is often not well covered in standard respiratory textbooks, but it certainly cannot replace these.—ARW

One of the nodular deposits of amyloid showing dichroism in polarised light. Stain: Congo red. Original magnification × 100, reduced to 75% in reproduction.