Z Rechtsmed (1990) 103:379–383

Zeitschrift für
Rechtsmedizin
© Springer-Verlag 1990

Casuistics

Sudden death due to bilateral spontaneous pneumothorax caused by rupture of congenital lung cysts

K. Simonsen

Institute of Forensic Medicine, Odense University, JB Winsløwsvej 17, DK-5000 Odense C, Denmark

Received August 29, 1989

Summary. A case of sudden death of a 35-year-old woman due to spontaneous bilateral pneumothorax from rupture of congenital lung cysts is reported. The woman had been attended by two doctors before the lethal outcome. The medicolegal aspects pertaining to medical negligence, the diagnostic difficulties and the rare occurrence of the condition are discussed, as is the autopsy procedure.

Key words: Sudden death – Bilateral pneumothorax – Congenital lung cysts – Autopsy procedure – Medicolegal aspects

Zusammenfassung. Berichtet wird über einen plötzlichen Todesfall einer 35 Jahre alten Frau, Tod durch spontanen bilateralen Pneumothorax, verursacht durch die Ruptur angeborener Lungenzysten. Vor dem tödlichen Ausgang war die Frau durch 2 Ärzte untersucht worden. Die verschiedenen rechtsmedizinischen Aspekte, wie Fahrlässigkeit im ärztlichen Beruf, diagnostische Schwierigkeiten, Seltenheit dieses Vorfalls, und auch das Procedere bei der Autopsie werden diskutiert.

Schlüsselwörter: Plötzlicher Tod – Bilateraler Pneumothorax – Kongenitale Lungenzysten – Obduktionstechnik – Rechtsmedizinische Aspekte

Introduction

Primary spontaneous pneumothorax is defined as a spontaneous pneumothorax in a patient in whom there are no clinical findings (other than the pneumothorax per se) referable to the underlying etiology of the spontaneous pneumothorax. This is in contrast to secondary spontaneous pneumothorax, which is defined as a spontaneous pneumothorax that occurs as a consequence of a manifest disease

process [1]. These definitions are somewhat arbitrary, as even primary spontaneous pneumothorax is probably always secondary to some underlying pathologic process. The important fact is, however, that the conditions referred to as primary spontaneous pneumothorax are unexpected and may thus be difficult to recognize. One case of sudden death caused by simultaneous bilateral primary spontaneous pneumothorax arising from rupture of congenital lung cysts is presented.

Case report

A 35-year-old white woman without any known previous pulmonary disease contacted her usual doctor by telephone on Friday 26th because of moderate respiratory distress. He attended her on the same day and found nothing abnormal on auscultation and percussion of both lungs. The patient had no dyspnea or cough, and the only abnormal finding was a distinct soreness of the muscles between the lower ribs on the right side. The doctor assumed that the patient's complaints were due to a muscle fiber sprain, and he gave a subcutaneous injection of pethidine (75 mg). On the morning of Monday 29th the woman called another doctor because of increased breathing difficulties. She explained that she had been well during the weekend but had suddenly become short of breath again that morning. This doctor found the woman dyspneic and pale, but not cyanotic. On auscultation of the lungs he found decreased breath sounds, but he found no differences between the two sides. He found nothing abnormal on percussion of the thorax. The examination revealed no indications of an asthmatic attack, and on the basis of the conversation with the woman he presumed that the condition was provoked by psychological factors. He gave her an injection of terbutalin, diazepam and morphine – all in therapeutic doses – and encouraged her to call again if her condition did not improve. A neighbor later explained to the police that 20 min after the doctor had left the woman's condition suddenly and rapidly worsened. An ambulance was called; by the time it arrived the woman was unconscious, and she was dead on arrival at the emergency room. Resuscitative measures were tried, including external cardiac massage, intubation, ventilation and intracardial adrenaline injection, all of which proved unsuccessful. When the first injection of adrenaline was given it was noted that air could be drawn into the syringe. As the death was sudden and unexpected and since the question of medical negligence was open, a medicolegal autopsy was ordered.

Autopsy findings

The thoracic cavities were incised at the base of a water-filled skin pocket. This technique revealed bilateral pneumothorax, and later massive atelectases of both lungs were demonstrated. In addition, several cavities were found at the apices of the lungs.

Toxicological analyses revealed therapeutic doses of diazepam and morphine in the blood, and examination for alcohol and other drugs was negative.

Histological examination of lung tissue revealed universal atelectases of the tissue except in samples from the apices where several cysts were demonstrated. Some of the cysts had no epithelial lining, while others were covered with a somewhat cubical epithelial cell layer. The adjacent tissue was fibrotic with old hemorrhages and calcifications, but no changes were found in the remaining lung tissue (Fig. 1).

It was concluded that the cause of death was bilateral spontaneous pneumothorax with bilateral atelectases of the lungs due to spontaneous rupture of congenital lung cysts in the lung apices. The case was later submitted to the National

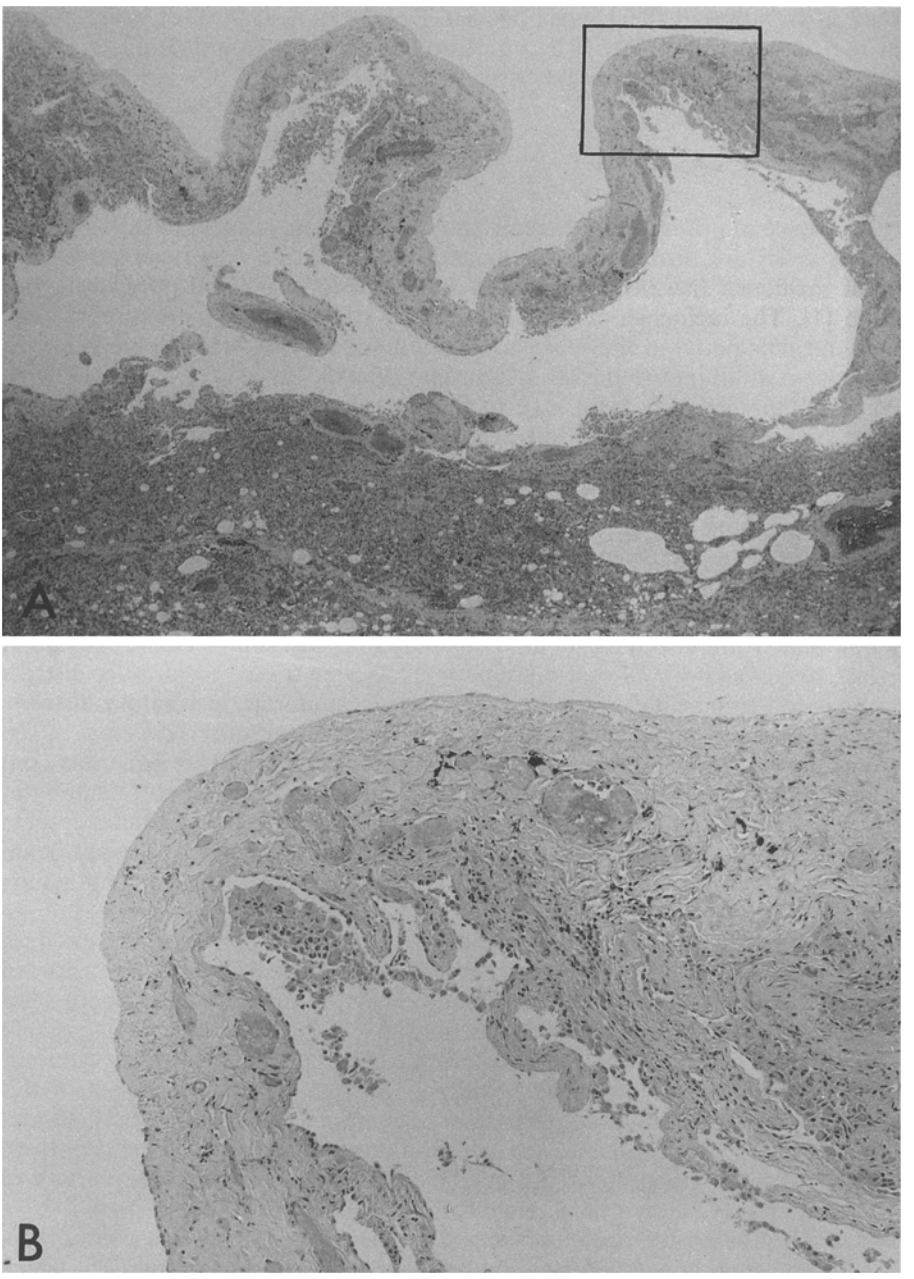

Fig. 1. A Large subpleural cysts and several smaller cysts adjacent to atelectatic lung tissue. H & E, × 24. **B** Cyst walls partly covered with cuboidal epithelial cells, some without epithelial lining. Note the fibrotic tissue with pigmentation from previous hemorrhages. H & E, × 105

Board of Health and the Danish Medico-Legal Council, both of which found that the two doctors involved had performed sufficient examinations and that the treatment given could not be criticized.

Discussion

The incidence of primary spontaneous pneumothorax is difficult to state exactly, as many cases are probably never diagnosed. In a large American survey the overall incidence has been calculated to be 7.6 per 100,000 population per annum [1]. The incidence of bilateral primary spontaneous pneumothorax has not been calculated, but there is usually simultaneous bilateral involvement in a few per cent of all reported cases of pneumothorax.

The author has only been able to find one reported case of sudden death caused by simultaneous bilateral primary spontaneous pneumothorax in the literature and it dates back to 1915 [2]. It was the case of a 19-year-old man with a past history of good health. The autopsy revealed several emphysemateous blebs in both lung apices. In this case it is not possible to determine for sure whether the blebs are congenital or acquired, but at that time it was concluded that they were acquired, probably as a consequence of previous tuberculosis.

Reports of cases of non-fatal simultaneous bilateral primary spontaneous pneumothorax are also infrequent. Thus only four cases have been reported in the last 10 years [3–6]. Two of these patients were in severe respiratory distress on admission to hospital; the other two were in moderate respiratory distress. All patients were treated successfully and made an uneventful recovery.

It is now believed that primary spontaneous pneumothorax generally, but not always develops as a result of rupture of one or more subpleural emphysematous blebs [1, 7]. In the present case the autopsy revealed congenital cysts in the apices of both lungs. It seems to be the first reported case of sudden death caused by bilateral spontaneous pneumothorax due to rupture of congenital lung cysts.

This patient probably first suffered from a right-sided pneumothorax as a result of rupture of one or more cysts. Subsequently – maybe because of forced respiration and increased pressure – additional ruptures of cysts on both sides have led to the fatal bilateral involvement.

Congenital cysts such as those in the present case probably result from arrested development of bronchial branching. Depending on the extent of this bronchial developmental arrest, the final appearance can be that of either congenital (saccular) bronchiectasis or congenital cystic disease. When very large numbers of cysts are present the condition is responsible for one variety of honeycomb lung seen in infants dying soon after birth [8].

As illustrated by the present case and previously reported cases, simultaneous bilateral spontaneous pneumothorax is a potentially fatal condition, which should be recognized and treated promptly. Furthermore, it is an extremely rare condition. It is also known that the diagnosis may be very difficult to make as the clinical features are varied and cannot be considered specific. In the present case the last doctor to examine the patient pointed out that he had found no differences between the two sides of the thorax on auscultation and percussion. In all cases an X-ray of the thorax should be performed to confirm or rule out the diagnosis of bilateral pneumothorax. The difficulties involved in recognition of the

condition are emphasized by the conclusion of both the National Board of Health and the Danish Medico-Legal Council that none of the doctors concerned could be criticized for their examination or treatment of the person in question.

From a medicolegal point of view it must be emphasized that examination for pneumothorax should be routinely performed in all medicolegal autopsies, since valuable information may otherwise be missed. Since the procedure is so easy this demand is reasonable.

Acknowledgements. The author thanks Mrs. S. Wandorf for skilful secretarial assistance and Assistant Professor E. Falk, MD, for help with micrographs.

References

1. Killen DA, Gobbel WG (1968) Spontaneous pneumothorax. Brown, Boston Little
2. Hayashi I (1915) Über tötlichen Pneumothorax durch Infarkt und Emphysem. Frank Z Pathol 16:1–71
3. Petterson GB, Gatzinsky P, Selin K (1983) A case of total bilateral spontaneous pneumothorax. Scand J Thorac Cardiovasc Surg 17:175
4. Raundahl U, Nielsen TH, Kristoffersen E, Dahl J (1986) Spontan bilateral total pneumothorax. Ugeskr Læger 148:2161
5. Gibb WRG, Maiwand MO (1986) Parietal pleurectomy for simultaneous bilateral spontaneous pneumothorax. Br J Clin Pract 40:489–490
6. Donovan PJ (1987) Bilateral spontaneous pneumothorax: a rare entity. Ann Emerg Med 16:1277–1280
7. Kjærgaard H (1932) Spontaneous pneumothorax in the apparently healthy. Acta Med Scand [Suppl] 43:1–59
8. Spencer H (1985) Pathology of the lung. Pergamon Press, Oxford New York, pp 98–102