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CASE FOR DIAGNOSIS

Cystic pulmonary lesion in a 6 year old girl

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Case report

A 6 year old German girl, with no history of any chronic disease, fell ill with cough and otitis. Penicillin treatment was started. Because of fever, abdominal pain and worsening of the coughing during the next days, a chest X-ray was performed. It showed an extensive infiltration of the right upper lobe. Under suspicion of pneumonia, oral antibiotic treatment was extended to amoxycillin for 10 days. Meanwhile, fever had decreased. However, chest X-ray showed no improvement. The girl was referred to our department for further diagnostic procedures.

Chest radiograph on admission (fig. 1) showed an infiltration of the right upper lobe, in which a fluid containing structure, with a round air bubble, attracted attention. In addition a dystelectasis of the middle lobe was noted. Because of these findings, the history and the subfebrile temperature of 38.5°C, an intravenous broad antibiotic treatment with fosfocine, gentamicin and metronidazole was started, under suspicion of an insufficiently treated pneumonia with liquefactive necrosis.

Laboratory data revealed an elevated erythrocyte sedimentation rate of 60/120 mm, C-reactive protein was 1.3 $mg \cdot l^{-1}$. Differential blood cell count was normal. Other data were also unremarkable: immunoglobulins, tumour markers alpha-fetoprotein (aFP) and human chorionic gonadotrophin (HCG), serological investigations concerning echinococcus and other pathogens. Intradermal tuberculin test with 10 tuberculin units (TU) purified protein derivative (PPD) remained negative.

For further diagnosis bronchoscopy was performed, which showed thick, purulent hypersecretion from the right upper lobe. The cystic lesion could not be reached by any instrument. Bacteriological investigations remained without growth of any micro-organism. Taking all findings into consideration, an unresolved abscess forming pneumonia seemed to be the most probable diagnosis, at that time, and thoracic surgical intervention was performed.

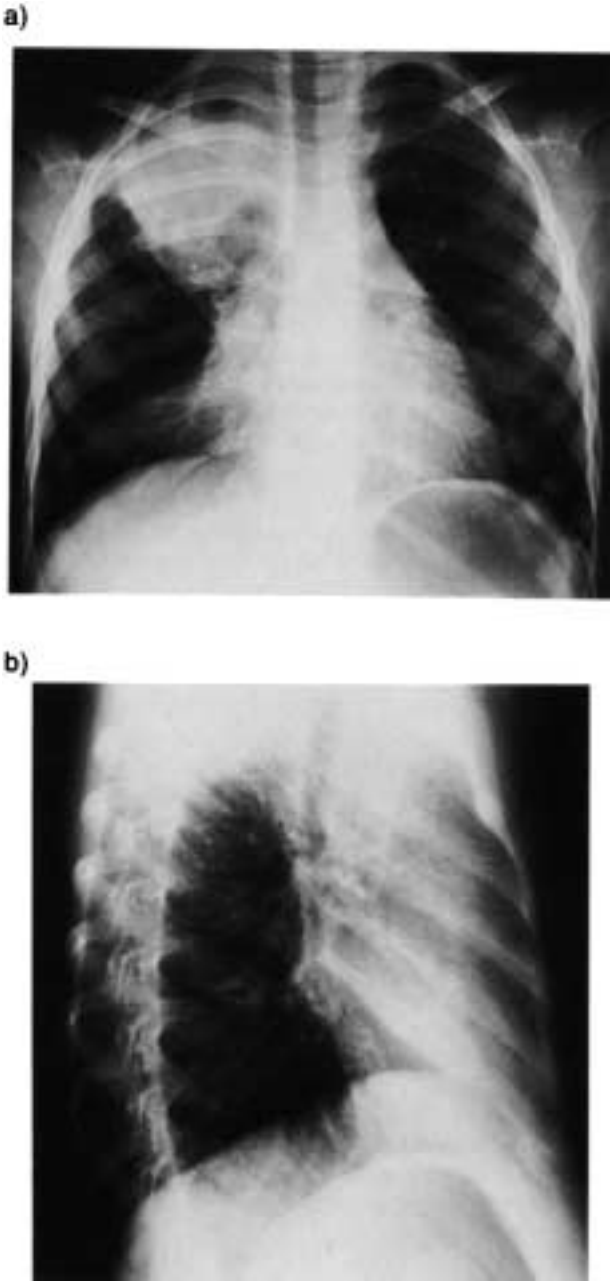

Fig. 1. – Chest radiograph on admission. a) posterioranterior view; b) lateral view.

TURN PAGE FOR DIAGNOSIS

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Intraoperatively, a cystic tumour 8 cm in diameter within the first and second segment of the right upper lobe was noted. A funnel-shaped configuration extended to the right hilar region. This part of the lung was supplied by two accessory blood vessels (artery and vein), which originated from the aortic arch and superior vena cava, respectively. An atypical resection by laser surgery was performed. No other abnormalities could be detected.

DIAGNOSIS: intralobar pulmonary sequestration of the right upper lobe

The histological work-up showed structures of an ectatic bronchus, fibrosis, and massive granulocyte infiltration. There were no signs of malignancy. After surgery, our patient recovered quickly, and no postoperative complications were seen. Chest radiographs weeks and months later showed a little remaining infiltration, which can be interpreted as scarred tissue.

Discussion

Pulmonary sequestration is a rare congenital malformation. In most cases a cystic mass of nonfunctioning lung tissue lacks normal communication with the bronchial tree, and a typical blood supply derives from systemic arteries [1]. There are two types of sequestration: intralobar (abnormal tissue and the normal lung are surrounded by the common visceral pleura), and extralobar (abnormal lung tissue is completely separated from normal lung tissue and with its own pleural investment). Intralobar sequestration is reported to be more common than extralobar [2–4], which is in keeping with our finding. Other authors found extralobar sequestration to be more common in their patients [5, 6].

As in our child, patients most often present clinically with unresolving pneumonia or recurrent chest infections [2, 7, 8]. Few asymptomatic children are diagnosed incidentally because of abnormal chest radiographs, performed for other reasons, but the chest X-ray is always abnormal [7, 8], and provides the main diagnostic clue. Cysts, as in our child, are a well-known sign of pulmonary sequestration in imaging techniques [3]. Arteriography can also provide proof. In our case, we did not perform an arteriogram, because unresolved abcedating pneumonia was thought to be a more probable diagnosis than pulmonary sequestration or an infected bronchogenic cyst [9].

A particular feature of our case report concerns localization: while the vast majority (98%) of cases of pulmonary sequestrations are found in the lung base [2, 8, 10], our child presented with striking localization of the right upper lobe. There are only a few other case reports with this localization in the literature [10, 11]. Arterial vascular supply in our child originated from the aortic arch, instead of the descending thoracic or abdominal aorta, typical for cases of pulmonary sequestration of the lung base [4].

Diagnosis is usually made in childhood, but there are case reports in adults also [12]. The management of pulmonary sequestration is thoracotomy, with tissue preserving resection of the abnormal lung mass, preferably after treatment of any intercurrent infection, as in our child.

In cases of unresolved or recurrent localized pneumonia, diagnosis of pulmonary sequestration should be considered, irrespective of localization within the lung.

Keywords: Bronchopulmonary; children; cyst; intralobular; lung; sequestration.

References

1. Pryce DM, Sellors TH, Blair LG. – Intralobular sequestration of lung association with an abnormal artery. *Br J Surg* 1917; 35: 18–29.
2. Allison RS, Chirnside AM. – Pulmonary sequestration: a review of 12 cases. *NZ Med J* 1983; 96: 381–383.
3. Ikezoe J, Murayam S, Godwin JD, Done SL, Verschakelen JA. – Bronchopulmonary sequestration: CT assessment. *Radiology* 1990; 176: 375–379.
4. John PR, Beasley SW, Mayne V. – Pulmonary sequestration and related congenital diorders. A clinicoradiological review of 41 cases. *Pediatr Radiol* 1989; 20: 4–9.
5. Buntain WL, Wooley MM, Mahour GH, Isaacs Jr H, Payne Jr V. – Pulmonary sequestration on children: a twenty five year experience. *Surgery* 1977; 81: 413–420.
6. de Paredes CG, Pierce WS, Johnson DG, Waldhausen JA. – Pulmonary sequestration in infants and children: a 20 year experience and review of the literature. *J Pediatr Surg* 1970; 5: 136–147.
7. Collin PP, Desjardins JG, Khan AH. – Pulmonary sequestration. *J Pediatr Surg* 1987; 22: 750–753.
8. Gustafson RA, Murray GF, Warden HE, Hill RC, Rozar GE. – Intralobar sequestration: a missed diagnosis. *Ann Thorac Surg* 1989; 47: 841–847.
9. Bailey PV, Tracy Jr T, Connors RH, de Mello D, Lewis JE, Weber TR. – Congenital bronchopulmonary malformations. Diagnostic and therapeutic considerations. *J Thorax Cardiovasc Surg* 1990; 99: 597–603.
10. Hoeffel JC, Bernard C. – Pulmonary sequestration of the upper lobe in children. *Radiology* 1986; 160: 513–514.
11. Gupta SK, Abraham KA, Ganesh KM. – Intralobular sequestration of upper lobe of right lung: case reports. *Angiology* 1988; 39: 1056–1060.
12. Sugio K, Kaneko S, Yokoyama H, Ishida T, Sugimachi K, Hasuo K. – Pulmonary sequestration in older children and in adults. *Int Surg* 1992; 77: 102–107.