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## **Endobronchial fibrosarcoma presenting as recurrent left-sided pneumonia.**

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## Endobronchial fibrosarcoma presenting as recurrent left-sided pneumonia.



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3 **Endobronchial fibrosarcoma presenting as recurrent left-sided pneumonia.**  
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## Summary

We report a six year old girl with recurrent and non-resolving left-sided 'pneumonia' over a three year period who was diagnosed with an endobronchial low-grade fibrosarcoma. This highlights the importance of considering underlying localised airway obstruction in any case of clinically or radiologically atypical pneumonia in a child and therefore the need for careful follow-up of all but the most typical cases.

## Introduction

Community-acquired pneumonia is relatively common in children, with an incidence of 10-40 cases per 1000 per year<sup>1</sup>. It is usually due to viral or bacterial infection in an otherwise healthy child and resolves completely<sup>1</sup>. Typically, such infections are clinically resolved before radiological resolution is complete<sup>2</sup>. When infections have an atypical course (such as recurrent episodes, incomplete resolution or atypical radiological appearances) or atypical features (for example failure to thrive, positive family history, gastrointestinal symptoms) further investigations are warranted to exclude diseases like Cystic Fibrosis, immunodeficiency, Primary Ciliary Dyskinesia and aspiration. We describe an unusual case of recurrent pneumonia which highlights two important lessons.

## Case

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3 A fully immunised six year old girl was referred to Royal Brompton Hospital for  
4 investigation of multiple episodes of a pneumonia-like illness over the previous three  
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8 years.  
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11 At three and a half years of age she had presented to the local hospital with a short  
12 history of fever, cough and vomiting. Chest radiography showed left-sided  
13 opacification. She was treated for community-acquired pneumonia with intravenous  
14 antibiotics and was discharged home on oral antibiotics after clinical improvement.  
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21 Ten days later she was readmitted with respiratory distress and fever. Chest  
22 radiograph showed persistent left sided opacification and was treated with 2 weeks of  
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26 broad spectrum intravenous antibiotics. Within three weeks of her recovery from this  
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31 episode, she was again admitted with respiratory exacerbation for intravenous  
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34 Follow up chest radiograph (two months after last episode of illness) showed some  
35 but not complete resolution of left sided opacification. Further investigations were  
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45 Three years later, at six and a half years of age, she presented again with intermittent  
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58 Clinical assessment showed dramatically reduced breath sounds throughout the left  
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3 concave to the left. Spirometry revealed an FEV1 of 0.82L, 67% predicted, FVC  
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5 1.05L, 79% predicted. Chest radiograph (*figure 1*) showed a collapse of the left lung  
6  
7 with a soft tissue opacity protruding into the left main bronchus. Contrast enhanced  
8  
9 chest CT scan (*figure 2*) confirmed obstruction of the left main bronchus by a mass.  
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11 Three small soft tissue nodules measuring 2-3mm were also noted in the right lung.  
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13 Flexible bronchoscopy (*figure 3*) revealed an endobronchial polypoid mass,  
14  
15 completely occluding the left main bronchus. Following biopsy confirmation of  
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17 neoplasia, classified at that time as low-grade spindle cell neoplasm not otherwise  
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19 specified, the patient underwent left pneumonectomy. At thoracotomy, the tumour  
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21 was found to be arising from a broad base within the left main bronchus and so a  
22  
23 sleeve resection was not possible. A left pneumonectomy was therefore performed  
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25 with resection of a firm broad-based mass measuring 30x25x20mm obstructing the  
26  
27 left main bronchus and involving the bronchial wall. The proximal bronchial resection  
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29 margin was examined by frozen section microscopy during the operation, and showed  
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31 no evidence of disease.  
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42 Microscopy showed a submucosal proliferation of mildly pleomorphic spindle cells  
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44 arranged in sheets, storiform whorls and fascicles with scattered multinucleate giant  
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46 cells and some peripheral inflammatory cells (*figure 4*). There were occasional  
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48 mitoses present but no necrosis. Staining for CD34 was very focally positive,  
49  
50 although positivity for this antibody was not viewed as specific for a tumour type in  
51  
52 this context. The tumour was negative for SMA and ALK-1, arguing strongly against  
53  
54 an inflammatory myofibroblastic tumour. Staining for MNF116 and EMA was  
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56 negative, thus excluding carcinoma from the diagnosis. Neural and perineural tumours  
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58 were excluded based on the lack of S-100 and claudin-1 positivity. There was no  
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3 histiocytic component morphologically and this was corroborated by the lack of  
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5 CD68 positivity within the neoplastic cells. PCR did not detect the FUS-CREB3L2 or  
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8 ETV6-NTRK3 gene fusions, thus ruling out a low grade fibromyxoid sarcoma and  
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10 congenial fibrosarcoma. Overall, the features were regarded as those of a low-grade  
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12 fibrosarcoma.  
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16 Serial follow-up chest CT scans (over last 2 years) have not shown any change in the  
17  
18 number, size or appearances of the nodules in the right lung.  
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## 20 21 22 **Discussion** 23

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26 Community-acquired pneumonia is a common in young children and usually resolves  
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28 completely with antibiotic treatment and supportive care<sup>1</sup>. In most cases it is usually  
29  
30 not essential to investigate for an underlying cause. In adults chest radiography is  
31  
32 routinely repeated after convalescence from community acquired pneumonia to screen  
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34 for underlying neoplastic disease<sup>3</sup>. Current guidelines on management of community  
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36 acquired pneumonia in children state that, “follow up chest radiography should only  
37  
38 be performed after lobar collapse, an apparent round pneumonia, or for continuing  
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40 symptoms<sup>1</sup>. This recommendation is supported by prospective studies of which  
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42 demonstrate no need for routine follow-up chest radiography<sup>2,4-6</sup>.  
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49 However, if there is a history suggesting progressive or persistent collapse or  
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51 consolidation it is important to consider the possibility, albeit rare, of a structural  
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53 abnormality including a neoplastic endobronchial lesion. Case series of endobronchial  
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55 neoplasms in children suggest that the majority of these present as persistent collapse  
56  
57 or consolidation<sup>7-10</sup> and that delay in diagnosis can have implications for future  
58  
59 surgical management.  
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3 Primary lung tumours in children are rare. They have a tendency to present  
4 endobronchially, in particular inflammatory myofibroblastic tumour, carcinoids,  
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6 salivary-gland tumours, hamartomas, papillomas and sarcomas. Inflammatory  
7  
8 myofibroblastic tumours are the most common primary pulmonary tumours in  
9  
10 children under the age of sixteen<sup>13</sup>. This entity was excluded in our patient based on  
11  
12 the lack of characteristic morphology, in particular an absence of inflammatory cells  
13  
14 within the tumour, and the absence of both SMA and ALK-1 staining. Guccion and  
15  
16 Rosen noted that the majority of endobronchial fibrosarcomas occurred in children,  
17  
18 whereas intrapulmonary fibrosarcomas presented mostly in adults and elderly  
19  
20 populations. They classified fibrosarcomas of the lung into endobronchial and  
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22 intrapulmonary types and this classification scheme was said to have prognostic and  
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24 clinical importance<sup>11</sup>. In another series, metastases were more common with  
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26 pleuropulmonary blastoma and carcinoid being more frequently seen than  
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28 mesenchymal neoplasms<sup>12</sup>.  
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## 45 Conclusion

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48 This child had recurrent consolidation in the left lung with several relapses despite  
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50 aggressive antibiotic treatment. She also never had a completely normal chest x-ray  
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52 and perhaps this should have prompted earlier referral and investigation. We have  
53  
54 reported this case because it illustrates two important lessons: firstly, the importance  
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56 of following up all but the most typical cases of community acquired pneumonia to  
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58 ensure complete recovery (clinical and radiological). Secondly, although rare, primary  
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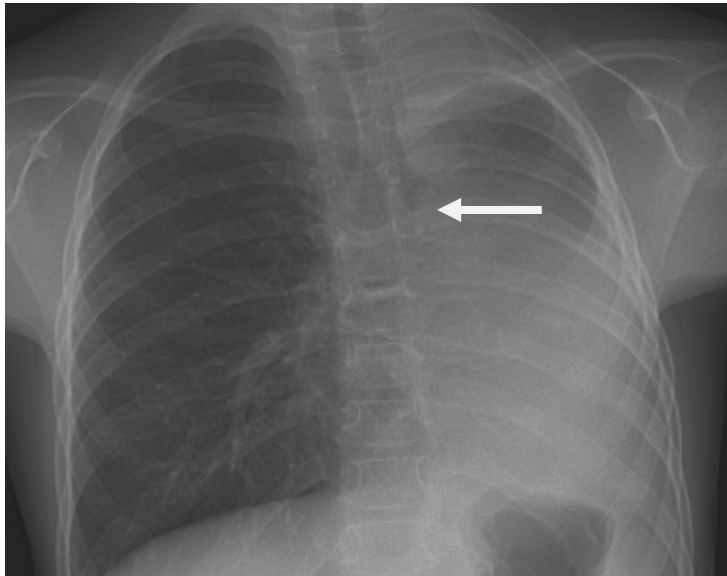
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pulmonary tumours can occur in children and should be considered in the differential diagnoses of non-resolving radiological shadowing.

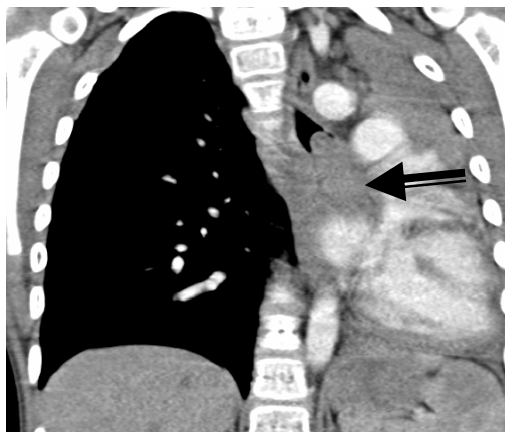
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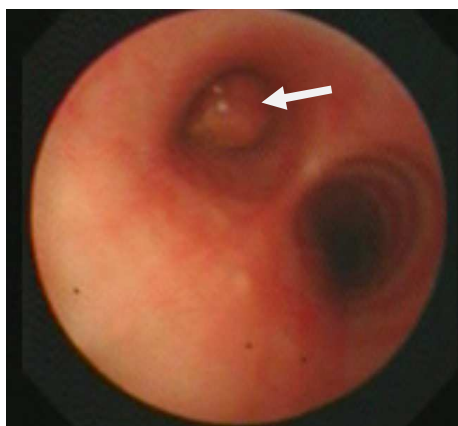
**Figure 1.** Chest x ray showing a collapse of the left lung with a soft tissue opacity, protruding into the left main bronchus.



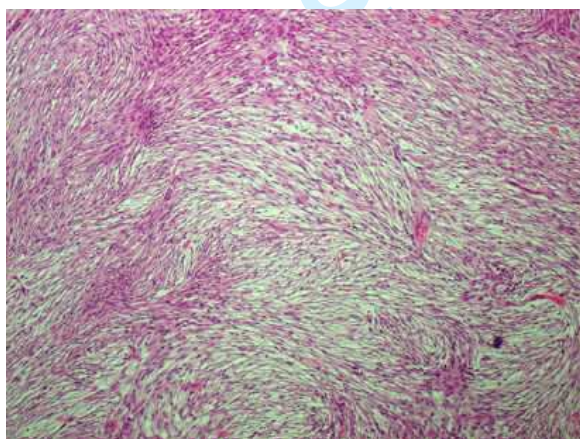
**Figure 2.** CT scan of chest confirmed a collapsed left lower lobe and a soft tissue mass in the left main bronchus extending into the left lung



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**Figure 3.** Flexible bronchoscopy showed a large endobronchial mass obstructing the left main bronchus



**Figure 4.** Microscopic image showing spindle cells arranged in storiform whorls and fascicles typical of a low-grade fibrosarcoma. (H&E x 100 magnification)

**References**

1. **British Thoracic Society Guidelines for the Management of Community Acquired Pneumonia in Childhood. Thorax 2002;57 Suppl 1:i1-24.**
2. **Virkki R, Juven T, Mertsola J, Ruuskanen O. Radiographic follow-up of pneumonia in children. Pediatr Pulmonol 2005;40(3):223-227.**
3. **Lim WS, Baudouin SV, George RC, Hill AT, Jamieson C, Le Jeune I, Macfarlane JT, Read RC, Roberts HJ, Levy ML, Wani M, Woodhead MA. BTS guidelines for the management of community acquired pneumonia in adults: update 2009. Thorax 2009;64 Suppl 3:iii1-55.**
4. **Heaton P, Arthur K. The utility of chest radiography in the follow-up of pneumonia. N Z Med J 1998;111(1072):315-317.**
5. **Gibson NA, Hollman AS, Paton JY. Value of radiological follow up of childhood pneumonia. BMJ 1993;307(6912):1117.**
6. **Suren P, Try K, Eriksson J, Khoshnewiszadeh B, Wathne KO. Radiographic follow-up of community-acquired pneumonia in children. Acta Paediatr 2008;97(1):46-50.**

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7. Fauroux B, Aynie V, Larroquet M, Boccon-Gibod L, Ducou le Pointe H, Tamalet A, Clement A. Carcinoid and mucoepidermoid bronchial tumours in children. *Eur J Pediatr* 2005;164(12):748-752.
8. Al-Qahtani AR, Di Lorenzo M, Yazbeck S. Endobronchial tumors in children: Institutional experience and literature review. *J Pediatr Surg* 2003;38(5):733-736.
9. Curtis JM, Lacey D, Smyth R, Carty H. Endobronchial tumours in childhood. *Eur J Radiol* 1998;29(1):11-20.
10. Hullo E, Llerena C, Durand C, Piolat C, Plantaz D, Pin I. [Recurrent pneumonia revealing a bronchial carcinoid tumor: report of two cases]. *Arch Pediatr* 2007;14(8):1036-1040.
11. Guccion JG, Rosen SH. Bronchopulmonary leiomyosarcoma and fibrosarcoma. A study of 32 cases and review of the literature. *Cancer* 1972;30(3):836-847.
12. Dishop MK, Kuruvilla S. Primary and metastatic lung tumors in the pediatric population: a review and 25-year experience at a large children's hospital. *Arch Pathol Lab Med* 2008;132(7):1079-1103.
13. Snyder CS, Dell'Aquila M, et al. Clonal changes in inflammatory pseudotumor of the lung. *Cancer* 1995;76:1545-9.

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Figure 1. Chest x ray showing a collapse of the left lung with a soft tissue opacity, protruding into the left main bronchus.  
96x75mm (72 x 72 DPI)

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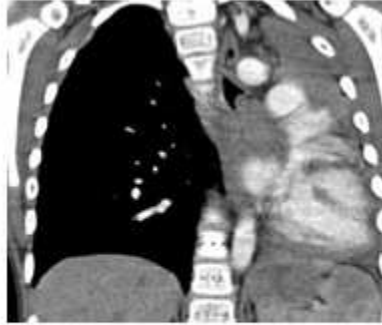


Figure 2. CT scan of chest confirmed a collapsed left lower lobe and a soft tissue mass in the left main bronchus extending into the left lung  
112x84mm (72 x 72 DPI)



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Figure 3. Flexible bronchoscopy showed a large endobronchial mass obstructing the left main bronchus  
59x55mm (72 x 72 DPI)

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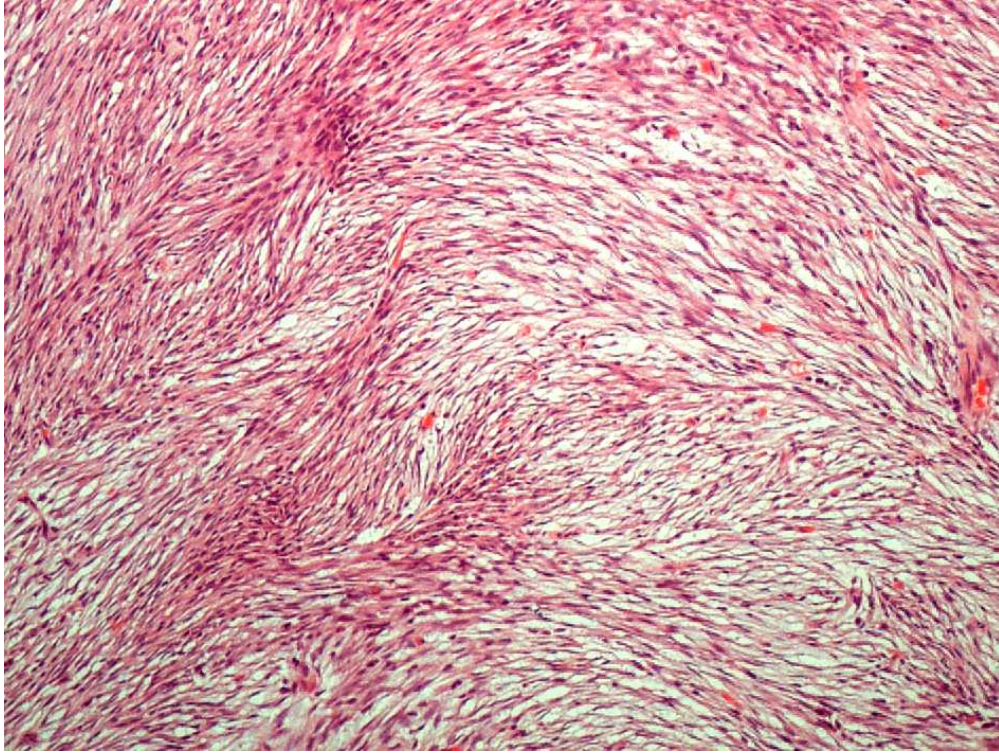


Figure 4. Microscopic image showing spindle cells arranged in storiform whorls and fascicles typical of a low-grade fibrosarcoma. (H&E x 100 magnification)  
80x60mm (300 x 300 DPI)

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8 **Authors** Gupta Atul<sup>1</sup>, Marsh Rosemary<sup>1</sup>, Jordan Simon<sup>2</sup>, Padley Simon<sup>3</sup>, Aboualfa  
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## Summary

We report a six year old girl with recurrent and non-resolving left-sided 'pneumonia' over a three year period who was diagnosed with an endobronchial low-grade fibrosarcoma. This highlights the importance of considering underlying localised airway obstruction in any case of clinically or radiologically atypical pneumonia in a child and therefore the need for careful follow-up of all but the most typical cases.

## Introduction

Community-acquired pneumonia is relatively common in children, with an incidence of 10-40 cases per 1000 per year<sup>1</sup>. It is usually due to viral or bacterial infection in an otherwise healthy child and resolves completely<sup>1</sup>. Typically, such infections are clinically resolved before radiological resolution is complete<sup>2</sup>. When infections have an atypical course (such as recurrent episodes, incomplete resolution or atypical radiological appearances) or atypical features (for example failure to thrive, positive family history, gastrointestinal symptoms) further investigations are warranted to exclude diseases like Cystic Fibrosis, immunodeficiency, Primary Ciliary Dyskinesia and aspiration. We describe an unusual case of recurrent pneumonia which highlights two important lessons.

## Case

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10 history of fever, cough and vomiting. Chest radiography showed left-sided  
11 opacification. She was treated for community-acquired pneumonia with intravenous  
12 antibiotics and was discharged home on oral antibiotics after clinical improvement.  
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15 radiograph showed persistent left sided opacification and was treated with 2 weeks of  
16 broad spectrum intravenous antibiotics. Within three weeks of her recovery from this  
17 episode, she was again admitted with respiratory exacerbation for intravenous  
18 antibiotics.  
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28 Follow up chest radiograph (two months after last episode of illness) showed some  
29 but not complete resolution of left sided opacification. Further investigations were  
30 done to exclude tuberculosis, Cystic Fibrosis and immune deficiency. Six months  
31 later she was discharged from follow-up after clinical resolution of her illness.  
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38 Three years later, at six and a half years of age, she presented again with intermittent  
39 dry cough for several weeks and short history of fever. She was treated with  
40 intravenous and oral antibiotics. Chest radiograph showed extensive left lung  
41 consolidation. After this episode she was referred to Royal Brompton Hospital for  
42 further management.  
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49 Clinical assessment showed dramatically reduced breath sounds throughout the left  
50 hemithorax with the apex beat displaced to the left axilla. There was a mild scoliosis  
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2 concave to the left. Spirometry revealed an FEV1 of 0.82L, 67% predicted, FVC  
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4 1.05L, 79% predicted. Chest radiograph (*figure 1*) showed a collapse of the left lung  
5  
6 with a soft tissue opacity protruding into the left main bronchus. Contrast enhanced  
7  
8 chest CT scan (*figure 2*) confirmed obstruction of the left main bronchus by a mass.  
9  
10 Three small soft tissue nodules measuring 2-3mm were also noted in the right lung.  
11  
12 Flexible bronchoscopy (*figure 3*) revealed an endobronchial polypoid mass,  
13  
14 completely occluding the left main bronchus. Following biopsy confirmation of  
15  
16 neoplasia, classified at that time as low-grade spindle cell neoplasm not otherwise  
17  
18 specified, the patient underwent left pneumonectomy. At thoracotomy, the tumour  
19  
20 was found to be arising from a broad base within the left main bronchus and so a  
21  
22 sleeve resection was not possible. A left pneumonectomy was therefore performed  
23  
24 with resection of a firm broad-based mass measuring 30x25x20mm obstructing the  
25  
26 left main bronchus and involving the bronchial wall. The proximal bronchial resection  
27  
28 margin was examined by frozen section microscopy during the operation, and showed  
29  
30 no evidence of disease.  
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35 Microscopy showed a submucosal proliferation of mildly pleomorphic spindle cells  
36  
37 arranged in sheets, storiform whorls and fascicles with scattered multinucleate giant  
38  
39 cells and some peripheral inflammatory cells (*figure 4*). There were occasional  
40  
41 mitoses present but no necrosis. Staining for CD34 was very focally positive,  
42  
43 although positivity for this antibody was not viewed as specific for a tumour type in  
44  
45 this context. The tumour was negative for SMA and ALK-1, arguing strongly against  
46  
47 an inflammatory myofibroblastic tumour. Staining for MNF116 and EMA was  
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49 negative, thus excluding carcinoma from the diagnosis. Neural and perineural tumours  
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51 were excluded based on the lack of S-100 and claudin-1 positivity. There was no  
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histiocytic component morphologically and this was corroborated by the lack of CD68 positivity within the neoplastic cells. PCR did not detect the FUS-CREB3L2 or ETV6-NTRK3 gene fusions, thus ruling out a low grade fibromyxoid sarcoma and congenital fibrosarcoma. Overall, the features were regarded as those of a low-grade fibrosarcoma.

Serial follow-up chest CT scans (over last 2 years) have not shown any change in the number, size or appearances of the nodules in the right lung.

### Discussion

Community-acquired pneumonia is a common in young children and usually resolves completely with antibiotic treatment and supportive care<sup>1</sup>. In most cases it is usually not essential to investigate for an underlying cause. In adults chest radiography is routinely repeated after convalescence from community acquired pneumonia to screen for underlying neoplastic disease<sup>3</sup>. Current guidelines on management of community acquired pneumonia in children state that, “follow up chest radiography should only be performed after lobar collapse, an apparent round pneumonia, or for continuing symptoms<sup>1</sup>. This recommendation is supported by prospective studies of which demonstrate no need for routine follow-up chest radiography<sup>2,4-6</sup>.

However, if there is a history suggesting progressive or persistent collapse or consolidation it is important to consider the possibility, albeit rare, of a structural abnormality including a neoplastic endobronchial lesion. Case series of endobronchial neoplasms in children suggest that the majority of these present as persistent collapse or consolidation<sup>7-10</sup> and that delay in diagnosis can have implications for future surgical management.

1  
2 Primary lung tumours in children are rare. They have a tendency to present  
3 endobronchially, in particular inflammatory myofibroblastic tumour, carcinoids,  
4 salivary-gland tumours, hamartomas, papillomas and sarcomas. Inflammatory  
5 myofibroblastic tumours are the most common primary pulmonary tumours in  
6 children under the age of sixteen<sup>13</sup>. This entity was excluded in our patient based on  
7 the lack of characteristic morphology, in particular an absence of inflammatory cells  
8 within the tumour, and the absence of both SMA and ALK-1 staining. Guccion and  
9 Rosen noted that the majority of endobronchial fibrosarcomas occurred in children,  
10 whereas intrapulmonary fibrosarcomas presented mostly in adults and elderly  
11 populations. They classified fibrosarcomas of the lung into endobronchial and  
12 intrapulmonary types and this classification scheme was said to have prognostic and  
13 clinical importance<sup>11</sup>. In another series, metastases were more common with  
14 pleuropulmonary blastoma and carcinoid being more frequently seen than  
15 mesenchymal neoplasms<sup>12</sup>.

## 36 37 **Conclusion**

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39  
40 This child had recurrent consolidation in the left lung with several relapses despite  
41 aggressive antibiotic treatment. She also never had a completely normal chest x-ray  
42 and perhaps this should have prompted earlier referral and investigation. We have  
43 reported this case because it illustrates two important lessons: firstly, the importance  
44 of following up all but the most typical cases of community acquired pneumonia to  
45 ensure complete recovery (clinical and radiological). Secondly, although rare, primary  
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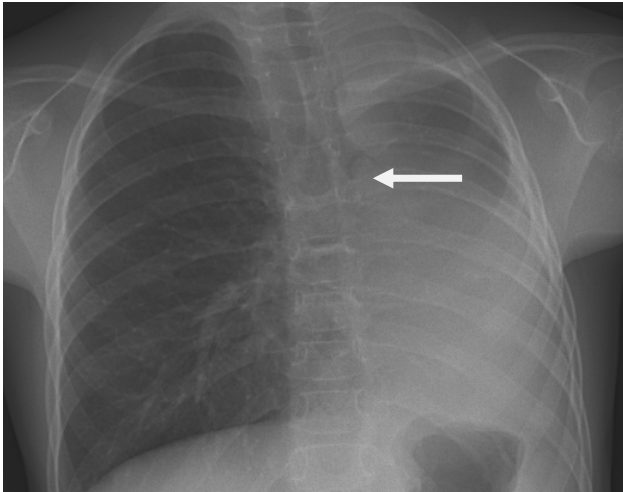


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pulmonary tumours can occur in children and should be considered in the differential diagnoses of non-resolving radiological shadowing.

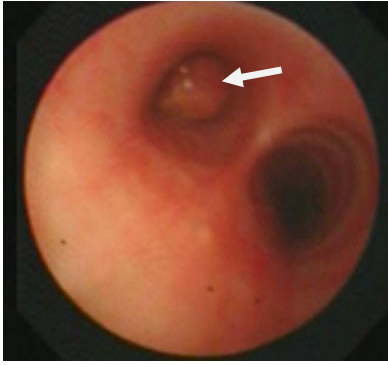
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3 **Figure 1.** Chest x ray showing a collapse of the left lung with a soft tissue opacity,  
4 protruding into the left main bronchus.  
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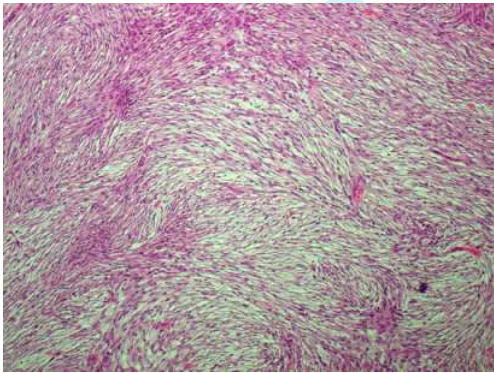


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26 **Figure 2.** CT scan of chest confirmed a collapsed left lower lobe and a soft tissue  
27 mass in the left main bronchus extending into the left lung  
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15 **Figure 3.** Flexible bronchoscopy showed a large endobronchial mass obstructing the  
16 left main bronchus



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33 **Figure 4.** Microscopic image showing spindle cells arranged in storiform whorls and  
34 fascicles typical of a low-grade fibrosarcoma. (H&E x 100 magnification)

## References

1. **British Thoracic Society Guidelines for the Management of Community Acquired Pneumonia in Childhood. Thorax 2002;57 Suppl 1:i1-24.**
2. **Virkki R, Juven T, Mertsola J, Ruuskanen O. Radiographic follow-up of pneumonia in children. Pediatr Pulmonol 2005;40(3):223-227.**
3. **Lim WS, Baudouin SV, George RC, Hill AT, Jamieson C, Le Jeune I, Macfarlane JT, Read RC, Roberts HJ, Levy ML, Wani M, Woodhead MA. BTS guidelines for the management of community acquired pneumonia in adults: update 2009. Thorax 2009;64 Suppl 3:iii1-55.**
4. **Heaton P, Arthur K. The utility of chest radiography in the follow-up of pneumonia. N Z Med J 1998;111(1072):315-317.**
5. **Gibson NA, Hollman AS, Paton JY. Value of radiological follow up of childhood pneumonia. BMJ 1993;307(6912):1117.**
6. **Suren P, Try K, Eriksson J, Khoshnewiszadeh B, Wathne KO. Radiographic follow-up of community-acquired pneumonia in children. Acta Paediatr 2008;97(1):46-50.**

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2  
3 7. Fauroux B, Aynie V, Larroquet M, Boccon-Gibod L, Ducou le Pointe H,  
4 Tamalet A, Clement A. Carcinoid and mucoepidermoid bronchial  
5 tumours in children. *Eur J Pediatr* 2005;164(12):748-752.  
6  
7
- 8  
9 8. Al-Qahtani AR, Di Lorenzo M, Yazbeck S. Endobronchial tumors in  
10 children: Institutional experience and literature review. *J Pediatr Surg*  
11 2003;38(5):733-736.  
12  
13
- 14  
15 9. Curtis JM, Lacey D, Smyth R, Carty H. Endobronchial tumours in  
16 childhood. *Eur J Radiol* 1998;29(1):11-20.  
17  
18
- 19  
20 10. Hullo E, Llerena C, Durand C, Piolat C, Plantaz D, Pin I. [Recurrent  
21 pneumonia revealing a bronchial carcinoid tumor: report of two cases].  
22 *Arch Pediatr* 2007;14(8):1036-1040.  
23  
24
- 25  
26 11. Guccion JG, Rosen SH. Bronchopulmonary leiomyosarcoma and  
27 fibrosarcoma. A study of 32 cases and review of the literature. *Cancer*  
28 1972;30(3):836-847.  
29  
30
- 31  
32 12. Dishop MK, Kuruvilla S. Primary and metastatic lung tumors in the  
33 pediatric population: a review and 25-year experience at a large  
34 children's hospital. *Arch Pathol Lab Med* 2008;132(7):1079-1103.  
35  
36
- 37  
38 13. Snyder CS, Dell'Aquila M, et al. Clonal changes in inflammatory  
39 pseudotumor of the lung. *Cancer* 1995;76:1545-9.  
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