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Mimics of Acute Pneumonia in Children and Adolescents: A Pictorial Review of Radiological Appearances

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Abstract

Respiratory symptoms commonly present in children and adolescents and, in most cases, are due to infection. Radiological imaging is generally reserved for cases in which there is sufficient diagnostic uncertainty or illness severity so as to justify the associated exposure to ionising radiation. Plain film radiography is the first line imaging modality, however computed tomography (CT), when performed in carefully selected cases, can more accurately evaluate thoracic anatomy and pathology and, in doing so, may reveal an alternative, sometimes unexpected, diagnosis.

This review serves to explore the variety of thoracic pathology that may precipitate presentation with respiratory symptoms in children and adolescents and, in doing so, to emphasise the importance of the differential diagnosis in children with an atypical presentation or disease course. In addition, it highlights the role of radiological imaging in carefully selected children, for whom detailed evaluation of thoracic anatomy and pathology can inform appropriate management and improve outcome.

INTRODUCTION

Community-acquired acute pneumonia due to viral (or, less commonly, bacterial) infection, is common in children. The diagnosis is generally made clinically, without radiological imaging. Indeed, studies have shown poor correlation between clinical signs and chest radiograph (CXR) findings, an inability to differentiate between bacterial or viral aetiology andno improvement in outcome (1).

If, however, a child is very unwell, first line treatment fails or there is high clinical suspicion of a separate disease process, imaging may be considered. If CXR does not answer the clinical question, alternative imaging, including computed tomography (CT), magnetic resonance (MR), ultrasound and/or fluoroscopy may be considered.

In this pictorial review, findings from thoracic CT of children and adolescents, performed over a 10-yearperiod at a tertiary centre, are presented, in order to demonstrate the variety of pathology that may occur in children, other than infection. Pathology presented ranges from the relatively common to the infrequent, from the acute to the chronic and from the reversible to the life-limiting.

CONGENITAL

Congenital Pulmonary Airway Malformation (CPAM)

CPAM is a type of bronchopulmonary foregut malformation, involving abnormal bronchial proliferation and cyst formation, and accounts for 25% of congenital lung lesions (2). The diagnosis is generally made antenatally. Rarely, CPAM is diagnosed during investigation of acute, progressive respiratory distress in a neonate. More rarely, the malformation is so small that the diagnosis is made later in life, as a consequence of recurrent chest infection (3).

CXR shows a multi-cystic lesion, usually involving a single lobe. In the neonate, cysts may be fluidfilled, appearing solid and larger lesions may cause mediastinal shift. CPAM may be mistaken for pneumonia, based on clinical and radiological findings and should be considered in cases of persistent/ recurrent consolidation in the same location (4). Once identified or suspected, CT enables differentiation between CPAM and pulmonary sequestration, as, unlike in the latter, there is no systemic arterial supply in CPAM (except in the setting of a so-called hybrid lesion, where both abnormalities are present).





Figure 1. Term baby presenting on day 1 of life with respiratory distress. (1a) CXR demonstrating left-sided thoracic cystic malformation with mediastinal shift.



Figure 1(b). Axial CT showing a multi-loculated, thickwalled mass arising from the lingula with no systemic blood supply in keeping with CPAM.

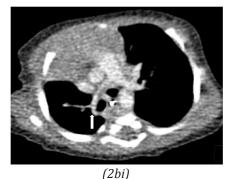
Hypogenetic Lung Syndrome

Hypogenetic lung (or Scimitar) syndrome is a subtype of congenital pulmonary venolobar syndrome, in which there is anomalous pulmonary venous return into the systemic circulation from a hypoplastic lung. Usually the right lung is involved, with drainage of either the middle or lower lobes (occasionally the whole lung) into the inferior vena cava (5). It can be present in association with other cardiopulmonary abnormalities thus presenting with signs of cardiac failure. In older children, symptoms are more variable and include recurrent respiratory infections and exertional breathlessness (6).

Appearances on CXR include a hypoplastic right lung with ipsilateral mediastinal shift and a tubular structure, adjacent tothe right heart border, representing the anomalous drainage vessel (and taking the shape of a Scimitar sword). CT or MR enable accurate delineation of vascular anatomy and evaluation of co-existing abnormalities.



(2a)





(2bii)

Figure 2. 8-day-old baby presenting with intermittent tachypnoea, poor feeding and faltering growth. (2a) CXR showing hyperinflation of the left lung with mediastinal shift to the right and loss of the right heart border. (2bi) Axial CT demonstrating a tubular structure communicating with the right-sided superior vena cava (white arrow) and inferior vena cava, with associated hypoplasia of the right lung and right pulmonary artery. In addition, there was absence of right pulmonary veins and right upper lobe bronchus. (2bii) Coronal CT image demonstrates anatomy described

Aberrant Right Subclavian Artery (ARSA)

ARSA (arteria lusoria) is the most common aortic arch abnormality, involving anomalous origin of the right subclavian artery, distal to the left subclavian

artery. Its course back to the right side most often involves running posteriorly to the oesophagus (7). Symptoms arise from extrinsic compression on the trachea or oesophagus. Infants may present with stridor or respiratory distress, prompting suspicion of infection. Indeed, respiratory infection is a recognised consequence of vascular rings in general (8).

CXR may demonstrate a mediastinal abnormality, however alternative imaging, e.g. fluoroscopy or CT, may be required in order to confirm the diagnosis and plan treatment.



Figure 3. 6-month-old baby presenting with coryzal symptoms and dyspnoea on a background of recurrent pneumonia. Initial CXR showed right lower zone consolidation which progressed over subsequent days. (3a) Barium swallow study showed posterior indentation of the upper oesophagus (black arrow) consistent with extrinsic compression from a vascular anomaly.

Congenital Diaphragmatic Hernia (CDH)

CDH (Bochdalek and Morgagni subtypes) cause morbidity and/or mortality due to resultant lung hypoplasia and derangement of the pulmonary vasculature and related structures (9). Large defects are usually detected during the antenatal period. Rarely, when small and undetected in the neonatal period, CDH may be found incidentally when imaging a child for a presumed pneumonia (10).

On CXR, fluid-filled or collapsed bowel loops will be homogenous and can mimic dense consolidation. The presence of an air-fluid levelmay make the diagnosis of CDH much easier, although necrotising pneumonia and lung abscess may also have this appearance on CXR. US and fluoroscopy can be of value.



Figure 4. Term baby presenting on day 2 of life with tachypnoea. (4a) CXR showing a large left diaphragmatic hernia containing small bowel and causing significant displacement of the mediastinum withcompression of both lungs.

Pulmonary Arteriovenous Malformation (PAVM)

In PAVM, abnormally dilated vessels provide a rightto-left shunt between the pulmonary artery and vein (11). PAVM may remain asymptomatic for some time (often detected during screening for hereditary haemorrhagic telangiectasia, with which there is an association), or present with respiratory distress (secondary to shunting and subsequent heart failure), embolic events or bleeding.

CXR may show a non-specific soft tissue mass (12). CT allows evaluation of PAVM number, size, nature, and suitability for embolisation and is preferred to MR because of greater resolution.



Figure 5. 6-year-old girl with mitochondrial disorder presenting with an upper gastro-intestinal bleed and respiratory distress. Initial CXR demonstrated bilateral airspace consolidation.(5a) Axial CT showing a large pseudo-aneurysm in the right upper lobe and a large AVM involving the mediastinum and extending into both lungs, fed by large bronchial arteries and multiple arterial branches arising from the thoracic aorta. There was shunting into the pulmonary venous system with pulmonary vein dilatation.

Congenital Pulmonary Agenesis

Pulmonary agenesis and aplasia result in unilateral or bilateral (not viable) absence of lung tissue. The two subtypes can be differentiated by the presence of the rudimentary blind-ending bronchus in pulmonary aplasia (13). Patients may initially be asymptomatic and present later in life with recurrent chest infections or, in more severe cases, present in the neonatal period with respiratory distress (14). The diagnosis needs to be considered in patients of any age who present with reduced breath sounds, no movement of unilateral chest wall and an opaque hemithorax on CXR. CXR may also reveal narrowing of the intercostal spaces, elevation of the diaphragm and mediastinal shift. (13).

CT or MR imaging is required to confirm the diagnosisand differentiate between pulmonary agenesis and aplasia (13).



(6a)



(6b)

Figure 6. Preterm baby presenting on day 1 of life in respiratory distress. (6a) Initial CXR demonstrating left-sided consolidation and right-sided collapse with mediastinal shift. (6b) Axial CT demonstrating absence of right lung, right pulmonary artery and right main bronchus, with complete shift of the mediastinum and consequent over-inflation of the left lung.

Inflammatory/Immunological

Aspiration

Aspiration pneumonia (related to food, saliva and/ or secretion) is particularly common in children with pre-existing diseases or disabilities(15). Discrimination between infective and aspiration pneumonia is difficult based solely on clinical findings and radiological appearances may be of value.

CXR shows consolidation in the dependent lung segments (the posterior segment of the upper lobes and the superior segment of the lower lobes), although this is relatively non-specific. CT features are more specific, with centrilobular and tree-in-bud nodules seen in aspiration pneumonia. Anterior lung involvement is also rarely reported in aspiration pneumonia. Water soluble swallow (under fluoroscopic guidance) +/- CT is usually required to establish any underlying mechanical problems or to identify tracheoesophageal or tracheopulmonary fistula(16).



Figure 7. Ex pre-term baby presenting at 8 months of life with recurrent chest infections and chronic cough. A barium swallow showed severe gastro-oesophageal reflux, with contrast reaching the cervical oesophagus. (7a)Axial CT demonstrating background features of chronic lung disease, as well as organising pneumonia in both lower lobes and the right middle lobe.

Granulomatosis with polyangitis (GPA)

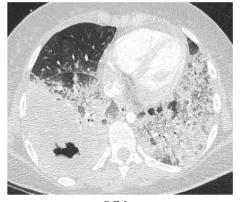
GPA is an ANCA-associated small-medium vessel vasculitis, associated with granulomatous inflammation, predominantly affecting the renal and respiratory tracts, which is rare in children (17).

Diagnostic criteria for childhood GPA includes radiological evidence of pulmonary involvement (present in 50 to 80% of patients). Features include multifocal lung nodules, which may be

cavitating, consolidation and ground glass opacities (GGO) (secondary to alveolar haemorrhage). GGO surrounding a nodule might lead to the "halo" sign (18). Differentiation from bronchopneumonia requires a high index of suspicion as radiological appearances are similar. Reactive lymphadenopathy is more common in infection.







8(b)

Figure 8. 12-year-old girl with 4-week history of cough, fatigue and fever. (8a) CXR showing cavitating right lower lobe mass (8b) Axial CT confirming large, thick-walled pulmonary nodule, with central cavitation and surrounding GGO. After inadequate response to treatment, further investigation confirmed raised anti-

PR3 antibodies, consistent with GPA.

Eosinophilic Pneumonia

Eosinophilic pneumonia, in which there is diffuse infiltration of airway and lung interstitium by eosinophils, with associated tissue injury, presents rarely in children (19). It can occur as a primary entity, or as secondary sequela of infection or neoplasia, or in response to drug exposure.

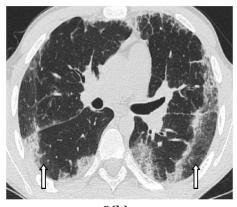
Given its rarity in children, it is difficult to define

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characteristic radiological features. In one retrospective review of five cases of primary or idiopathic eosinophilic pneumonia, radiography showed bilateral peripheral alveolar infiltration with ill-defined margins in almost all cases.CT might also demonstrate diffuse GGO and reticulonodular densities(20).



9(a)



9(b)

Figure 9. A 16-year-old boy presented with recurrent lower respiratory tract infection and persistently raised serum eosinophils (9a) CXR showing bilateral peripheral infiltrates (9b) Axial CT showingbilateral, asymmetrical, peripheral GGO with interlobular septal thickening (white arrows). Biochemical markers, along with the clinical picture and the radiology, allowed for a diagnosis of eosinophilic pneumonia.

Sarcoidosis

Sarcoidosis is a multi-system disorder, pathologically defined by granulomatous inflammation. It predominates in young adults, in which thoracic involvement is almost universal. Incidence in children is low and thoracic involvement less likely in those under 5 years of age (21).

CT enables parenchymal evaluation, withfeatures including mediastinal lymphadenopathy, pulmonary nodules, GGO and interlobular septal thickening. Contrast-enhanced MRI using fast sequences may be comparable to CT in evaluating thoracic sarcoidosis, while avoiding the use of ionising radiation(22).

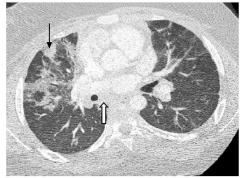


Figure 10. 10-year-old boy presenting with progressive dyspnoea and intermittent small-volume haemoptysis. (10a) Axial CT showing evidence of widespread, noncavitating ground glass nodules (black arrow) with hilar and mediastinal lymph node enlargement (white arrow). A diagnosis of sarcoidosis was subsequently made.

Children's Interstitial Lung Disease (chILD)

Neuroendocrine cell hyperplasia of infancy (NEHI)

NEHI is a rare interstitial lung disease of childhood that typically presents as respiratory distress in the first year of life and isassociated with significant morbidity and mortality(23).

CXRshows marked hyperinflation, increased interstitial markings and mild peri-hilar opacification. CT typically demonstrates GGO (with a central, middle lobe and lingula predominance) and air trapping with a mosaic pattern (24). If inconclusive, lung biopsy can be performed.







11(b)

Figure 11. Four-month-old baby presenting with increased work of breathing. (11a) CXR demonstrating perihilar airspace shadowing and consolidation in the right lower zone(11b) Axial CT chest demonstrating hyperinflated lungs with bilateral GGO involving predominantly the upper lobes, lingula segment and right middle lobe, radiological findings were enough to confirm a diagnosis of NEHI.

Surfactant Metabolism Dysfunction

Surfactant deficiency and associated respiratory distress syndrome (RDS) in preterm infants is well recognised and, as such, all preterm infants receive surfactant replacement. A less well understood cohort of patients are term babies with inborn errors of surfactant metabolism, who present with RDS and subsequently develop chILD(25, 26).



Figure 12. Term baby presenting with respiratory distress and failure to thrive. Initial CXR demonstrated non-specific findings with bilateral GGO and hyperinflation. (12a) Axial CT demonstrating overinflated lungs with GGO, peripheral hyperlucency with areas of mosaicism and a marked pectus deformity. Genetic testing confirmed an ABCA3 mutation in keeping with surfactant metabolism dysfuction.

Imaging findings are non-specific and genetic testing +/- biopsy is required for a diagnosis. CXR typically demonstrates bilateral diffuse or patchy opacities and CT findings include GGO, septal thickening, parenchymal cysts and pectus excavatum (25, 26).

Cardiac

Atrial Septal Defect

Cardiac failure can present with dyspnoea, poor feeding and poor growth during infancy. Older children and teenagers may complain of fatigue, especially if there is myocardial damage. Cardiac failure can clinically mimic pneumonia or other respiratory infections. CXR might show cardiomegaly and there can be increased pulmonary blood flow in left to right shunts, with enlarged pulmonary vessels, seen more peripherally. Increased pulmonary venous pressure suggests left-sided cardiac dysfunction, and the margins are indistinct as interstitial oedema occurs (27). On CT, this manifests as septal thickening. Subtle GGO can be seen in a dependent distribution with a HU difference of 100-150 between the dependent and non-dependent lung. Bilateral pleural fluid can be seen. An increase in size of the aortic knuckle suggests secondary increased blood flow (e.g. PDA, truncus arteriosus, valvular insufficiency) or decrease in aortic knuckle size due to decreased blood flow (e.g. ASD, VSD or hypoplastic left heart syndrome).

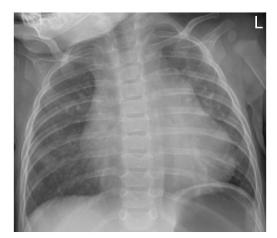


Figure 13. 6-month-old male presenting with progressive dyspnoea. (13a) CXR demonstrated increased bronchovascular markings with left lower lobe consolidation. The heart size is difficult to assess on this AP projection. The patient subsequently had an echocardiogram which demonstrated a large ASD.

Pulmonary Malignancy

It is uncommon for a pulmonary malignant mass lesion to be diagnosed incidentally on a CXR(28). Pulmonary metastases are uncommon in children and it is rarer still to develop primary tumours. In thoracic neuroblastic tumours, picked up co-incidentally on CXR, a paraspinal mass can be seen associated with posterior rib destruction or separation. Conversely, round pneumonia in a child with an infective history should not be misinterpreted as a neoplastic lesion.

Lymphoma

There are four patterns in the radiographic appearances of pulmonary lymphoma in children; nodular, alveolar, lymphangitic or miliary nodules (29). The most common finding is multiple nodules with air bronchograms (30). The halo sign can also be demonstrated on CT due to localised infiltration of lymphoma cells into the surrounding lung tissue or secondary to invasion of lymphoma cells into blood vessels with subsequent bleeding.

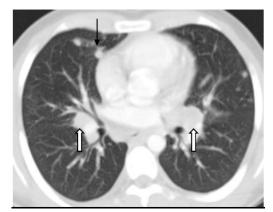


Figure 14. 15-year-old boy presenting with a cough. (14a) Axial CT demonstrated bilateral lymph nodes, with surrounding ground glass change (black arrow). Enlarged mediastinal lymph nodes (white arrows) are also seen. A diagnosis of lymphoma was subsequently made.

Metastases

Metastatic lung disease is rare but most commonly from osteosarcoma, which is associated with calcified lung lesions. More common causes include Ewing's sarcoma, rhabdomyosarcoma, hepatoblastoma and Wilms tumours. CT is more sensitive in detection of pulmonary metastatic disease compared to radiographic imaging.

CONCLUSION

While pneumonia is common, when there is a high index of suspicion of an alternative diagnosis, radiological imaging can support diagnosis of a number of conditions that may present with non-specific cardio-respiratory symptoms. Diagnoses range from congenital anatomical variants, amenable to definitive surgical management, to chronic conditions, requiring long-term management. The radiologist role is to support selection of appropriate imaging, taking into account the likelihood of a diagnostic imaging study based on the most likely differentials, as well as the risks of ionising radiation.

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