Original article

Chest radiographic findings in children with asplenia syndrome

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Background: Early diagnosis of asplenia syndrome is important because prophylactic antibiotic and proper vaccination will prevent serious infection. Most children with asplenia syndrome present with symptoms of congenital heart disease. Chest X-ray is usually the first line imaging modality.

Objective: Define useful findings in chest radiograph that could suggest the diagnosis of asplenia syndrome.

Methods: Chest radiographs of pediatric patients who had asplenia syndrome diagnosed between January 1, 2002 and June 30, 2008 in a single institute were retrospectively reviewed for the positions of the visceral organs in the chest and abdomen.

Results: Three hundred seventy one chest radiographs of 30 patients were reviewed. The mean age at diagnosis was 3 years old. Asplenia was diagnosed by ultrasound in 27 patients, by CT scan in two patients, and by damaged red blood cell scintigraphy in one patient. Six important findings detected from chest radiographs were, 1) bilateral minor fissures, 16 cases (53%), 2) bilateral eparterial bronchi, 16 cases (53%), 3) ipsilateral side of stomach and liver, 12 cases (40%), 4) ipsilateral side of minor fissure or eparterial bronchus and stomach, 10 cases (33%), 5) symmetrical transverse lie of the liver, nine cases (30%), and 6) contralateral side of minor fissure or eparterial bronchus and the liver in seven cases (23%). All except two patients (93%) had at least one of the above findings. All patients had congenital heart diseases. Most of the heart diseases were pulmonary atresia or pulmonary stenosis (88%) and single ventricle (85%).

Conclusion: Chest radiographs have high sensitivity in suggesting the diagnosis of asplenia syndrome, when detecting one or more of the above findings, particular in patients with congenital heart disease and decreased pulmonary vasculature.

Keywords: Asplenia, chest X-ray, children, congenital heart disease, heterotaxia, minor fissure, situs ambiguous

Asplenia syndrome is one of the sub-classifications of heterotaxy syndrome or situs ambiguous [1, 2]. It has other names such as bilateral right-sidedness and right isomerism. Bilateral presence of organs and structures that are normally found on the right side and absence of organs such as spleen, which are normally found on the left side. Patients with asplenia syndrome typically have bilateral tri-

lobed lungs, bilateral eparterial bronchi, transverse lie of the liver, and no spleen [1, 3-5].

Asplenia is often associated with severe complex cyanotic congenital heart disease (CHD) [6-8] with high mortality rate in the first year of life [6]. Better surgical techniques result in better post-operative outcomes and longer life expectancy [9]. Apart from CHD, the other significant abnormality is the life-long risk of infections because of the lack of splenic macrophages and asplenic patients make fewer antibodies [10, 11]. Early diagnosis of asplenia syndrome will lower the risk of serious infection, because antibiotic prophylaxis and appropriate vaccination [12] can be given.

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Plain chest radiography is commonly obtained in patients with CHD and in patients with immune deficiency. In this study, we reviewed chest radiographs of asplenic patients to look for findings that may give clues in suggesting the diagnosis of asplenia syndrome.

Materials and methods

We retrospectively reviewed the medical records and imaging studies of patients with asplenia syndrome aged between 1 day and 15 years at King Chulalongkorn Memorial Hospital between January 1, 2002 and June 30, 2008. Absence of the spleen was determined by either ultrasonography (US), CT scan, damaged red blood cell scintigraphy, or at laparotomy. Patients were excluded if they had history of splenectomy or their chest radiographs were not available. All chest radiographs (both pre- and post-operative) of each subject were reviewed by one radiologist who had 9-year experience in pediatric radiology.

The structures to be evaluated with chest radiograph were, 1) position of the cardiac apex (levocardia, mesocardia, or dextrocardia), 2) cardiac size (normal or enlarged), 3) position of the aortic arch (right sided, left sided, or undetermined), 4) pulmonary vascularity (increased, normal, or decreased; symmetry or asymmetry), 5) bronchus (bilateral eparterial bronchi or not), 6) minor fissure (right only, left only, bilaterally present, or bilaterally not seen), 7) position of the liver (right, left, or symmetrically transverse), and 8) position of the stomach (right, mid, or left).

Suspicion of situs ambiguous with asplenia syndrome from chest radiographs were determined by one or more of the following findings, 1) bilateral minor fissures, 2) bilateral eparterial bronchi, 3)

symmetrical transverse lie of the liver, 4) ipsilateral side of stomach and liver, 5) ipsilateral side of minor fissure or eparterial bronchus and the stomach, 6) contralateral side of minor fissure or eparterial bronchus and the liver. These findings are usually not found in situs solitus or situs inversus. However some of them (symmetrical transverse liver, and ipsilateral side of stomach and liver) could be found in situs ambiguous with polysplenia syndrome.

Results

Thirty patients with asplenia syndrome that had plain chest radiographs and medical records were included in this study. All chest radiographs of these 30 patients were reviewed, thus, 371 films (range 2-48 films per person). Sixteen of the studied population was boy (53%). The mean age at diagnosis was three years (range: six days to 14 years). The absence of spleen was determined by US in 27 patients, CT scan in two patients, and by damaged red blood cell scintigraphy in one patient. None was diagnosed from laparotomy.

All patients had CHD. Enough information of cardiac disease from medical record was available in 26 patients (**Table 1**). The three most common cardiac anomalies were single ventricle (85%), atrial situs ambiguous, and common atrioventricular valve. Right ventricular outflow tract obstruction was very frequent. Pulmonary stenosis or atresia was found in 88%.

Pulmonary vascularity from chest radiograph was assessed in 25 patients, excluding four patients whose available chest radiographs were all post operative, and another one patient with persistent perihilar pneumonia and atelectasis. Pulmonary vascularity was decreased in 68%. Findings from chest radiographs were summarized in **Table 2**.

Table 1. Cardiovascular anomalies in 26 patients.

Cardiovascular anomalies	Cases (percent)
Single ventricle	22 (85%)
Atrial situs ambiguous	17 (65%)
Common AV-valve	14 (54%)
Pulmonary stenosis	13 (50%)
Pulmonary atresia	10 (38%)
Common atrium	8 (31%)
ASD primum without common AV-valve	3 (11%)

Table 2. Radiographic findings in 30 asplenic patients.

Findings	Cases	Findings	Cases
<i>Positions of cardiac apex</i>		<i>Minor fissure</i>	
Levocardia	17	Bilaterally	16
Mesocardia	1	Right only	8
Dextrocardia	12	Left only	4
		Both unseen	2
<i>Cardiac size</i>		<i>Eparterial bronchi</i>	
Normal size	18	Bilaterally	16
Cardiomegaly	12	Right only	6
		Left only	2
		Both indeterminate	6
<i>Position of aortic arch</i>		<i>Liver</i>	
Left arch	18	Symmetrical transverse	9
Right arch	10	More on the right	11
Undetermined arch	2	More on the left	9
		Indeterminate	1
<i>Pulmonary vascularity</i>		<i>Stomach</i>	
(Excluded)	(5)	Right	18
Decreased	17	Left	11
Normal	5	Mid	1
Increased	3		

The minor fissures could be detected on both sides in 16 patients (53%) (**Fig. 1**), right side only in eight, left side only in four, and could not be seen on either side in two. There was one patient who had persistent right upper lobe atelectasis with elevated minor fissure, we categorized this case in the group of right minor

fissure only. Minor fissures were more commonly seen in early post-operative period, when there were some degree of pulmonary congestion and pleural effusion.

Bilateral eparterial bronchi were seen in 16 patients (53%) (**Fig. 2**).

Fig. 1 Bilateral minor fissures (arrows) in a child with dextrocardia.

Fig. 2 Bilateral eparterial bronchi. Symmetrical and early branching of both right and left upper lobe bronchi were indicated by double arrows in chest radiograph (A), and by single arrow in three-dimensional image of virtual CT bronchography in another patient (B). T= trachea.

For the position of upper abdominal viscera, the hepatic shadow was found on the right side in 11 cases, left side in nine cases, and symmetrical transverse lie in nine cases (**Fig. 3**). In one patient, there was generalized bowel ileus in every film and the inferior edge of the liver was unclear.

Our predictors of suspecting situs ambiguous with asplenia syndrome from chest radiographs were one or more of 1) bilateral minor fissures, 2) bilateral eparterial bronchi, 3) symmetrical transverse lie of the liver, 4) ipsilateral side of stomach and liver, 5) ipsilateral side of minor fissure or eparterial bronchus and stomach, 6) contralateral side of minor fissure or eparterial bronchus and the liver. One score was given for each positive finding. The maximal score was three, because if one predictor was positive, some of the others would not, such as when 3) transverse lie of the liver was positive, there would be no score for 4) or 6).

Using the predictors as shown in **Table 3**, we found that 16 patients had bilateral minor fissures (53%) (**Fig. 1**), 16 had bilateral eparterial bronchi (53%) (**Fig. 2**), nine patients had transverse lie of the liver (30%) (**Fig. 3**). Twelve patients had ipsilateral side of liver and stomach (40%) (**Fig. 4**), 10 patients had ipsilateral side of minor fissure or eparterial bronchus and stomach (33%) (**Fig. 5**), and six patients had contralateral side of minor fissure or eparterial bronchus and liver (**Fig. 6**).

Twenty-eight patients (93%) had at least one of the above findings, 25 patients (83%) had at least two findings, and 17 patients (57%) had three findings. The suspected diagnosis of asplenia syndrome, using a single predictor, was usually made within the first eight films (**Fig. 7**). In 14 patients (47%), asplenia syndrome could be suspected on their first chest radiographs using any of the criteria.

Fig. 3 Symmetrical transverse lie of the liver in two patients (**A** and **B**).

Table 3. Predictors of asplenia syndrome from chest radiograph.

Predictors	Cases
Bilateral minor fissures	16 (53%)
Bilateral eparterial bronchi	16 (53%)
Symmetrical transverse liver	9 (30%)
Ipsilateral side of liver and stomach	12 (40%)
Ipsilateral side of minor fissure or eparterial bronchus and stomach	10 (33%)
Contralateral side of minor fissure or eparterial bronchus and liver	7 (23%)

Fig. 4 The liver and stomach were on the same right side. Dextrocardia and right sided aortic arch were noted. There was asymmetrical pulmonary vasculature, decreased on the right side, and increased on the left from the left modified Blalock Taussig shunt.

Fig. 5 The minor fissure and the stomach were seen on the same right side.

Fig. 6 The minor fissure and the liver were on opposite sides. In one child (A), well-expanded right upper lobe with normal position of right minor fissure (arrow) was noted, while in another patient (B), there was the right upper lobe atelectasis and the minor fissure (double arrows) was elevated.

Fig. 7 The relationship between the numbers of chest films (per person) to be evaluated and percentage of patients when a single radiographic criteria of asplenia syndrome was detected.

Discussion

All asplenic patients diagnosed in our institution came for medical care because of complex CHD. Our data showed that the most common cardiac diagnosis was single ventricle (85%), followed by atrial situs ambiguous (65%) and atrioventricular canal defect (either complete atrioventricular canal or ASD primum) (65%). In some previous reports [2, 6], atrioventricular canal defect had been described as the most common congenital heart malformation in asplenia syndrome.

Pulmonary atresia or severe stenosis were very common (88%) and in some cases there were additional unilateral pulmonary arterial stenosis. This explained why plain radiographs in six patients showed asymmetrically decreased pulmonary vascularity. Obstruction of the ventricular exit was reported to be found in about 98% [2].

Cardiomegaly, dextrocardia, right sided aortic arch, and decreased pulmonary vasculature were commonly found from chest X-ray of patients with asplenia syndrome, but these findings were not specific and could not be used as a clue for suggesting the diagnosis.

More specific findings found in asplenia syndrome were 1) bilateral minor fissures, 2) bilateral eparterial

bronchi, 3) transverse lie of the liver, 4) ipsilateral side of the liver and the stomach, 5) ipsilateral side of minor fissure or eparterial bronchs and the stomach, and 6) contralateral side of minor fissure or eparterial bronchus and the liver. By using the above six findings, all except two of our 30 patients showed at least one suspicious finding from chest radiographs.

“Bilateral minor fissures” sign was very useful in our experience, and were easy to be detected. We found bilateral minor fissures in 53% of our asplenic patients. The right and the left minor fissures might not be detected on the same film, so looking in every previous film would increase sensitivity in finding bilateral minor fissures. It was more frequently detected in early post thoracic operation, because of some degree of pulmonary congestion and pleural effusion. However, false positive can occur. Left minor fissure can be found from frontal view of chest radiograph with the incidence of 1.6% in normal population [13].

Detection of bilateral eparterial bronchi was challenging. Bronchial pattern is neither constantly nor easily seen in normal infants and young children. What we looked for was symmetrical early branching of right and left upper lobe bronchi. We succeeded in detecting bilateral eparterial bronchi in 53% of our

patients. Displaced lobar bronchus or displaced segmental bronchus of upper lobe arising more proximally can mimic eparterial bronchus of right isomerism, and can cause false positive diagnosis.

When only unilateral minor fissure or unilateral eparterial bronchus was found, the suspicion of asplenia syndrome still could be made if we saw the gastric fundus (by seeing gastric air or gastric tube) located on the same side, or found the liver on the opposite side.

Symmetrical transverse lie of the hepatic shadow were found in nine in 30 cases (30%).

If the liver was more located on one side, and we found the ipsilateral side of the stomach, situs ambiguous was also considered, and it was detected in 12 cases (40%) in this study. There were some limitations when assessing position of the liver. Transverse liver or ipsilateral side of liver and stomach could be found in polysplenia syndrome as well. Transverse liver in normal population had also been found. Besides, precise position and alignment of the liver might be difficult to evaluate when there was less air or too much air in bowel loops.

Position of the stomach alone was not helpful in suggesting the diagnosis of situs ambiguous.

The largest series of X-ray in asplenia was in 1973 by Freedom and Fellows [14] who evaluated apparent visceral situs from chest and abdominal radiographs of 32 asplenic patients using positions of the liver and stomach as the criteria. The pattern resembling situs ambiguous (horizontal liver, or a stomach bubble and a liver mass on the same side) was found in 50% of cases. They recognized eparterial bronchi in a careful review, but did not mention of its prevalence. Applegate et al. [7] studied chest radiographs in 11 asplenic children in 1999, and showed lower prevalence of bilateral minor fissures and bilateral eparterial bronchi comparing to ours, 3% vs. 53% and 1% vs. 53%, respectively. This was probably due to more recent technology of film-screen and the X-ray machine used in our patients, particular the use of computed radiograph in about one-third of our cases, which allowed adjustment of density and contrast during the review. Besides we reviewed every film of each patient and combined the result, so the detection rate of bilateral tri-lobed lung might be higher. The prevalence of midline horizontal liver and of the ipsilateral side of the liver and the stomach were not much different between Applegate's and our

reports, 45% vs. 30%, and 36% vs. 40%, respectively.

The retrospective study by itself had limitations and pitfalls. Most of our cases were initially suspected of having asplenia by chest radiographs, so the incidence of positive findings from chest X-ray may be high. The radiologist knew the diagnosis of asplenia before the review, which might result in over-emphasis of bronchial pattern or minor fissure even when they were very subtle, which might not be applicable in daily routine reading of X-ray films. Transverse lie of the liver with mild asymmetry was also subjective and may be affected by bias. If a prospective study is planned to review chest radiographs in all children with complex congenital heart disease, the incidence of positive findings from chest X-ray may be different.

However, from our data, we believed that plain chest radiographs were very helpful in suspecting asplenia syndrome, but it was necessary to evaluate every film carefully. The positive findings from chest radiographs would lead to further investigation to confirm the diagnosis. US, damaged RBC nuclear scintigraphy, CT scan, and magnetic resonance imaging are comparable tools in making the definite diagnosis, but the former two are mostly used [7]. In our institution, US was preferred because of its easy access, high accuracy, and no radiation. Early diagnosis of asplenia syndrome will allow early antibiotic prophylaxis and appropriate vaccination to be given timely, resulting in lessening the patient's risk of serious infection.

Conclusion

Plain chest radiographs were very helpful in detecting the findings that could suggest the diagnosis asplenia syndrome. Detection of bilateral minor fissures, bilateral eparterial bronchi, symmetrical transverse lie of the liver, same side of minor fissure or eparterial bronchus and the stomach, and opposite side of minor fissure or eparterial bronchus and the liver, which were not the findings of situs solitus or situs inversus, were highly suggestive of situs ambiguous with asplenia syndrome. Single or combination of these findings should arouse the suspicion of asplenia syndrome, particular in pediatrics with complex CHD and decreased pulmonary vasculature.

The authors have no conflict of interest to declare.

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