

Case Report

Sail sign, significance in paediatrics and review of literature

Menon Narayanankutty Sunilkumar

*Associate Professor, Department of Pediatrics, Amala Institute of Medical Sciences, Amala Nagar, Thrissur, Kerala, India

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ABSTRACT

A sail-like, triangular projection from the mediastinum in chest radiographs is encountered in healthy children in the first few years of life. This has been proved to be the shadow of the thymus and is not of pathological significance. As it resembles a "sail" it is known as the thymic Sail Sign (SS). Literature review reported it is seen in about 10% of normal children. Interestingly many treating doctors are not aware about the significance of the SS and can be missed easily or not given proper importance. SS is also seen in conditions like pneumomediastinum (PNM), DiGeorge syndrome (DiGs) and others. The chance of SS conditions should be searched in every infant or children to achieve better outcome and to prevent complications. This case study report SS in an infant and highlight the SS with a review of literature.

Key words: Thymus gland, Chest radiograph, Sail sign, Pneumomediastinum, DiGeorge Syndrome

INTRODUCTION

A number of interesting and often amusing terms in Radiological terminology are intended to help us to understand and remember the pathological abnormality. The Sail Sign (SS), which is actually a sail-like appearance in the chest X-ray, is intriguing and is also important for diagnosis in children under pathological conditions. In children, SS is a triangular projection from the mediastinum reflecting the shadow of the normal thymus [1]. Radiographs of chests of the healthy children in the first few years of life may show this significant finding which is not pathological [2]. There are conditions such as left lower lobe collapse, pneumomediastinum (PNM) and DiGeorge syndrome (DiGs) are also associated with SS, but with different causes. These conditions can be diagnosed by a combination of physical examination and confirmatory chest radiograph, along with various recognized signs that are characteristic for each particular setting. This article reports a case of SS in 3 month old infant.

CASE REPORT

A 3 month old male infant was admitted in the ICU with breathing difficulty since the previous night. He was seen by a local doctor and treated for symptoms of nose block and low grade fever for the last 3 days. He was breathing spontaneously with no cyanosis. He was exclusively breastfed and there was no history of

Figure 1: Classic thymic sail sign (white arrow) and Thymic wave sign (black arrows)



any aspiration of breast milk or oral secretions. On general physical examination, baby was alert and active. He was febrile 100.6°F, heart rate 132/minute, respiratory rate 66/minute, weight 4.36 kg. anterior fontanelle soft and flat, skin with good colour and perfusion. His oxygen saturation was 100% throughout. His respiratory effort remained good. Birth history revealed that he was born to a 24 year old mother at 38 weeks gestation via spontaneous vaginal delivery weighing 3.9 kg with normal apgar score. His parents were of non-consanguineously married. His newborn period was normal. A quick

glucose check was 175 mg/dl. Chest radiograph showed no cardiomegaly with mildly prominent vascular markings and the characteristic SS was seen (Figure 1). A diagnosis of bronchiolitis was considered and the infant was placed under humidified oxygen. His general condition improved with symptomatic treatment given for fever with paracetamol drops. An echocardiogram showed normal chamber sizes and contractility. Hemogram revealed Hb (11 g/dl), PCV (33.7%), total count (10000/ μ l), neutrophils (47%), lymphocytes (48%), eosinophils (2.3%), monocytes (2.5%), basophils (0.2%), ESR (31mm/hr), and platelets count (210000/ μ l) were all found to be normal. Urine routine analysis indicated albumin (nil), sugar (nil), pus cells (1-2/hpf), epithelial cells (+/hpf), bile salt (negative), bile pigment (negative). Blood and urine culture sensitivity were normal the patient was discharged after full recovery. Serum Na^+ 133 meq/l, K^+ 4.4 meq/l, Cl^- 91 meq/l, bicarbonate 26 meq/l, glucose 144 mg/dl, total Ca^{++} 9.0 mg/dl. He was discharged on day 3 with advice to have proper breast feeding practices, avoid early weaning, immunize regularly and follow-up.

DISCUSSION

The daily routine practice of a Paediatrician will meet with many chest radiographs. In infants, the thymus is very large organ relative to the thoracic cavity; hence its radiographic shadow may be mistaken for cardiomegaly or pulmonary infiltrate. The thymus continues to grow until puberty, when it atrophies [2]. The normal newborn chest has a trapezoidal shape in antero-posterior (AP) films. On normal inspiration, the hemidiaphragms in an AP chest film are at the level of the 6th rib anteriorly and the 8/9th rib posteriorly. The lungs are more radiolucent than those of older children. Air bronchograms may be seen in the medial thirds of the lungs [1]. The heart is more spherical in shape than in an older child. The cardiothoracic ratio of newborns has a wide range of about 0.65 as upper limit [1], beyond which cardiomegaly should be considered. The thymus, in the antero-superior mediastinum, is usually seen in newborn which has right and left lobes. The borders of the heart and thymus mainly the inferior border may blend, or there may be a notch at the junction of the border of the heart with the inferior border of the thymus. Furthermore, the thymic shadow makes the aorta and pulmonary arteries difficult to identify. Sometimes, the thymus has an inferior margin with an acute angle which produces a SS (Figure 1). If there is rotation in films of patients with a SS, the thymus may appear shifted laterally, which may be incorrectly interpreted

as upper lobe pneumonia [3]. Occasionally, gentle undulation is seen as a result of adjacent rib compression along the entire lateral edge of the thymus which is termed the 'wavy thymic sign' (Figure 1) [1,3].

In our baby with respiratory distress, the chest radiograph showed normally aerated lungs and a bulging thymic lobe over the right hemithorax. This is the classic "thymic sail sign" [5], a frequently encountered radiographic finding in pediatrics, representing the triangular extension of the normal thymus out laterally (Figure. 1). The thymic contour is convex and the inferior border is straight, giving it a sail-like appearance. However, pneumonia of any lobe would not present as a mediastinum-based opacity associated to the horizontal inferior border [4,5]. The classic thymic sail sign (white arrow) and thymic wave sign (black arrows) are mentioned here.

The thymic sail sign is a normal finding in infants and should not be confused with the "spinnaker-sail sign" where the thymic lobe shadow is laterally and superiorly displaced due to PNM, as though an actual boat sail blown by strong wind [4,7]. The normal thymus has a soft texture and may present an undulation of its contour on radiographs caused by the ribs, known as the "thymic wave sign" (black arrows) [5]. This appearance helps to exclude mediastinal expansive masses such as teratomas which would not have a horizontal inferior border and may present with calcifications, in contrast to the normal thymus with soft tissue density [4,5].

Overwhelming studies proved too little attention to SS [3-6]. The treating doctor also should have an idea of the importance of the lateral chest radiograph. In the lateral view of the newborn, space anterior to the heart is filled by the thymus, whereas in adults this space should be filled with lung tissue and so it is lucent. Obliteration of this space in an adult indicates the presence of right ventricular enlargement or a mediastinal mass. In newborns, this space should be filled with a tissue density (i.e. the thymus). If this space is filled with air, it is indicative of a PNM and if this space is filled with lung tissue, it is indicative of thymic aplasia or hypoplasia as in DiGs [1,3].

PNM is secondary to alveolar rupture with dissection of air along the pulmonary interstitium into the mediastinum and usually causes little or no respiratory distress. The classic configuration of PNM is caused by elevation of the thymic lobes by air loculated within the anterior mediastinum.[8]. The

spinnaker sign (also known as the angel wing sign) is a sign of PNM seen on neonatal chest radiographs and refers to the thymus being outlined by air with each lobe displaced laterally and appearing like spinnaker sails. This is distinct from the SS appearance of the normal thymus where free mediastinal air lifts the thymus off of the heart and major vessels[7,8]. 'Rocker-Bottom Thymus ' **a new sign of PNM in the neonate also has been mentioned[9].** The occurrence of PNM and pneumothorax in the neonate and infants is relatively common[7-11] and so the knowledge of these signs are pertinent to not miss this dangerous condition and life saving measures initiated.

Thymus is a primary immune organ involved in T cell differentiation and thus cell mediated immunity. DiGs is an immunodeficiency syndrome characterized by hypoparathyroidism and T-lymphocyte deficiency due to thymic aplasia or hypoplasia. The thymus and the parathyroid glands both arise from the same pharyngeal pouches (3rd and 4th) during embryogenesis [12]. Failure of these pouches to develop results in deficiencies of the thymus and parathyroids. Patients with DiGs are susceptible to opportunistic infections and typically present initially with tetany and hypocalcemia (resembling seizures) due to hypoparathyroidism long before any immunodeficiency is appreciated. Other clinical features of DiGs include congenital heart and aortic defects, hypoplastic mandible, defective ears[12,13]. Febrile seizures are most common and are not associated with any electrolyte or calcium abnormalities. In this child, the serum calcium levels were normal and did not have any findings similar to a case of DiGs.

The role of imaging is best understood with an appreciation of the embryology, anatomy, and pathology of the thymus [14,15]. In this study, the chest radiograph was the main tool in our hands to pick up SS in this child. Lee et al prospectively screened radiographs of 10,238 patients to determine the incidence of the SS found in adults in their 40s or older. And the cause of the SS was assessed using computer tomography (CT).The incidence of SS on adult chest radiographs in their study was about 0.1% [16]. This case study concludes that awareness of SS among the paediatricians and treating physicians is very important in preventing unnecessary worries and also not missing serious life saving conditions.

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Corresponding Author:

Dr. M N Sunil Kumar, MD,DNB
Associate Professor,
Department of Pediatrics,
Amala Institute of Medical Sciences,
Amala Nagar,
Thrissur-680 555, Kerala, India
E-mail: sunilsree99@gmail.com

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