A Hyperlucent Hemithorax on a Chest Radiograph: Poland Syndrome as an Uncommon Extrapulmonic Source

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In an era of powerful imaging techniques, the conventional thoracic radiography has not lost its relevance as an accessible and inexpensive investigational modality. The diagnostic value that a plane chest X-ray may offer cannot be underestimated.

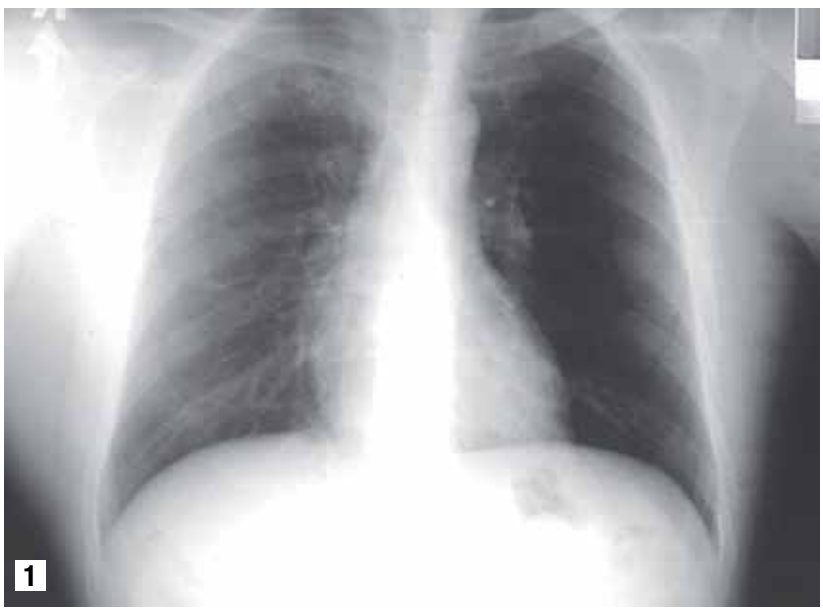
A 36 year old man presented with a few days history of episodic dry cough without fever. The never-smoking construction worker denied any history of thoracic trauma or previous surgery but had been aware of a chest wall irregularity since childhood. Family history was unremarkable. On pulmonary auscultation he had clear lungs with normal breathing and heart sounds.

A postero-anterior chest radiograph was interpreted as demonstrating diffusely hypodense left hemithorax and hyperlucent left lung without evidence of hyperinflation. The right lung and mediastinum were within normal limits [Figure 1].

Extended physical examination revealed an asymmetric chest, flattening of the left chest wall, and thinning of the left axillary fold due to aplasia of the sternocostal head of the left pectoralis major muscle, and elevated left nipple and areola [Figure 2]. The entire ipsilateral upper limb was hypoplastic, and the upper arm had a relatively smaller circumference with a hypoplastic biceps muscle. Examination of the hands disclosed a simple and incomplete syndactyly with short fingers establishing the diagnosis of Poland syndrome.

Poland syndrome is a congenital anomaly classically consisting of the combination of unilateral aplasia of the sternocostal head of the pectoralis major muscle and ipsilateral hypoplastic hand with simple syndactyly and short fingers [1]. Other associated anomalies may include ipsilateral rib anomalies such as hypoplasia or absence of ribs, and mammary tissue hypoplasia or aplasia [2]. The syndrome was first described in a cadaver by Sir Alfred Poland in 1841 after a few earlier reports [3]. The reported incidence of Poland syndrome ranges from 1 in 7000 to 1 in 100,000 live births [4]. Although most cases arise sporadically, genetic transmission also occurs. Sporadic cases tend to occur more often in males and commonly involve the right side. Familial cases have a more equal distribution between the sexes and do not have a right-sided predominance [3]. Currently the prevailing theory in the etiology of Poland syndrome is that at the end of the sixth week of gestation, when the upper limb bud adjacent to the chest wall is still in a stage of development, the interruption of embryonic blood supply causes hypoplasia of the ipsilateral subclavian artery or one of its branches [4].

A hyperlucent thorax on plain chest radiography may be attributed to both intrapulmonary conditions (obstructive or compensatory hyperinflation, pulmonary vascular disorder) and extrapulmonary causes (mastectomy, pneumothorax, contralateral pleural effusion, previous chest wall surgery, and suboptimal technique during radiography acquisition such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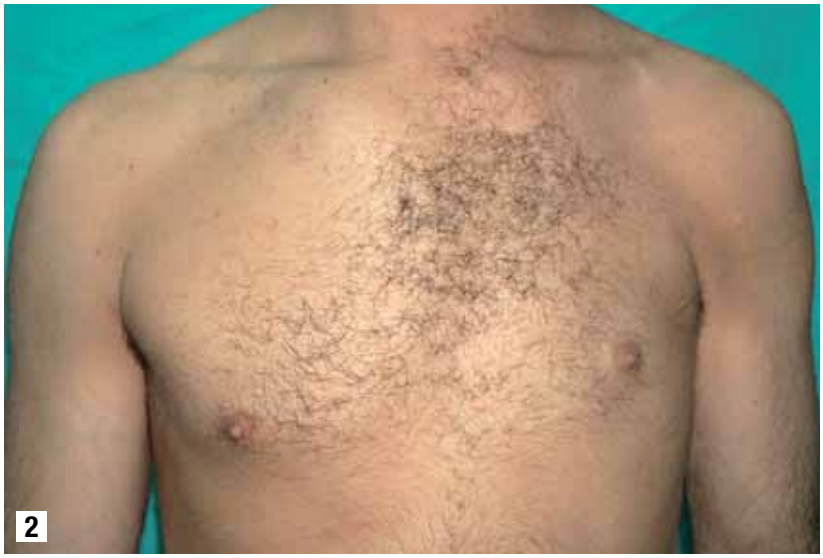

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as asymmetric positioning of the patient against the screen).

Poland syndrome should always be considered in the differential diagnosis of an apparently hyperlucent lung on radiography. An abnormal silhouette of the soft tissues may be helpful in identifying hypoplastic or absent muscles indicating an extrathoracic cause of hyperlucency. Anamnesis should be properly provided to both the X-ray technician and the radiologist for facilitating adequate interpretation.

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Capsule

Common and specific signatures of gene expression and protein–protein interactions in autoimmune diseases

In an attempt to understand intracellular regulatory mechanisms in peripheral blood mononuclear cells (PBMCs), which are either common to many autoimmune diseases or specific to some of them, Tuller et al. incorporated large-scale data such as protein–protein interactions, gene expression and demographic information of hundreds of patients and healthy subjects, related to six autoimmune diseases with available large-scale gene expression measurements: multiple sclerosis, systemic lupus erythematosus, juvenile rheumatoid arthritis, Crohn's disease, ulcerative colitis and type 1 diabetes. These data were analyzed concurrently by statistical and systems biology approaches tailored for this purpose. The authors found that chemokines such as CXCL1-3, 5, 6 and the interleukin-8 tend to be differentially expressed in PBMCs of patients with the analyzed autoimmune diseases. In addition, the anti-apoptotic gene *BCL3*, interferon-gamma, and the vitamin D receptor (*VDR*) gene physically interact with significantly many genes that tend to be differentially expressed in PBMCs of patients with the analyzed autoimmune diseases. In general, similar cellular processes tend to be differentially expressed in PBMC in the analyzed autoimmune diseases. Specifically, the cellular processes related to cell proliferation (for example, epidermal growth factor, platelet-derived growth factor, nuclear factor-κB, Wnt/β-catenin signaling, stress-activated protein kinase c-Jun NH2-terminal kinase), inflammatory response (for example, interleukins 2 and 6, the cytokine granulocyte-macrophage colony-stimulating factor and the B cell receptor), general signaling cascades (for example, mitogen-activated protein kinase, extracellular signal-regulated kinase, p38 and TRK) and apoptosis are activated in most of the analyzed autoimmune diseases. However, these results suggest that in each of the analyzed diseases, apoptosis and chemotaxis are activated via different subsignaling pathways. Analyses of the expression levels of dozens of genes and the protein–protein interactions among them demonstrated that Crohn's disease and ulcerative colitis have relatively similar gene expression signatures, whereas the gene expression signatures of type 1 diabetes and juvenile rheumatoid arthritis relatively differ from the signatures of the other autoimmune diseases. These diseases are the only ones activated via the Fcε pathway. The relevant genes and pathways reported in this study may be helpful in the diagnoses and understanding of autoimmunity and/or specific autoimmune diseases.

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