

Left sided Poland's syndrome associated with dextrocardia

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ABSTRACT

Poland's syndrome is a rare congenital anomaly coexisting with the absence of pectoralis major and ipsilateral upper limb and chest wall deformities. Hypoplasia of the breast, agenesis of ipsilateral rib cartilage, athelia, and an ipsilateral developmental finger anomalies as syndactyly also can be seen. In the literature, only 56 patients with dextrocardia and left-sided Poland's syndrome have been described. Herein, a case of left sided Poland's syndrome coexisting with dextrocardia and nasal hemangioma was presented.

Keywords: Dextrocardia; Poland's syndrome; nasal hemangioma.

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The Poland's syndrome is a rare congenital anomaly, first described by Sir Alfred Poland in 1941, characterized by unilateral agenesis of pectoralis muscles and ipsilateral hypoplasia of the breast, partial rib agenesis or hypoplasia and brachysyndactyly [1]. Hypoplasia or aplasia of serratus anterior muscle, latissimus dorsi muscle, and external oblique muscle have also been described. Poland's syndrome mostly involves the right side of the chest wall. Associated anomalies of Poland's syndrome are reported to be lung herniation, renal aplasia or hypoplasia, vertebral malformations, Moebius syndrome, frontonasal dysplasia and malignancies [2]. Although most cases of Poland's syndrome are sporadic, some authors reported cases with autosomal dominant and multifactorial inheritance [3]. The exact etiology of Poland's syndrome is unknown. Many theories have been suggested such as genetic factors, teratogenic effects and interruption of the subclavian artery during the early embryonic life [2]. The incidence of this syndrome has been reported to be 1/20.000–50.000 [4]. Poland's syn-

drome is more common in boys with a male-to-female ratio of 3:1 and more commonly affects the right side of the chest (67–75%) [5]. Coexistence of dextrocardia has rarely been reported in the literature and when present, dextrocardia is almost always associated with left-sided deformities. Herein, we presented a rare case of left sided Poland's syndrome associated with dextrocardia.

CASE REPORT

A 2-month-old male baby was admitted with complaints of congenital hemangioma on the tip of the nose (Figure 1). He was the sixth children of consanguineous marriage. His weight was 4,9 kg (25–50th percentile), length was 57 cm (25–50th percentile), occi 478/pitofrontal circumference was 39 cm (50–75th percentile). Initial physical examination revealed a depression of the left anterior chest wall, cranially located left nipple, hypoplastic right areola and hemifacial microsomia on the left side. In chest X-ray, dextrocardia and left-sided chest wall deformities were

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FIGURE 1. A 2-month-old male baby with congenital hemangioma on the nose, long filtrum and thin upper lip.

detected (Figure 2). Transthoracic echocardiography showed dextrocardia with suspicious connection of right pulmonary veins. Computed tomography (CT) of the chest was performed to exclude anomalies of pulmonary vein connections. Axial chest CT confirmed dextrocardia and demonstrated that right heart chambers were positioned posteriorly, left heart chambers anteriorly and descending aorta located on the left with normally connected vascular structures. There was asymmetry of right sided pectoralis muscles (white arrow) (Figure 3). Hypoplasia of left sided anterior ribs was demonstrated with 3D volume rendered CT (Figure 4). Hypoplasia of the left pectoralis muscle was confirmed by ultrasonography (Figure 5). Renal and testes ultrasonographic examination excluded the anomalies that can occur in Poland's syndrome such as renal aplasia or hypoplasia and undescended testes. Further systemic evaluation, including examination of hands, lower limbs, hair and nails did not show other anomalies. Neurological examination and transcranial ultrasonography were normal. In his familial

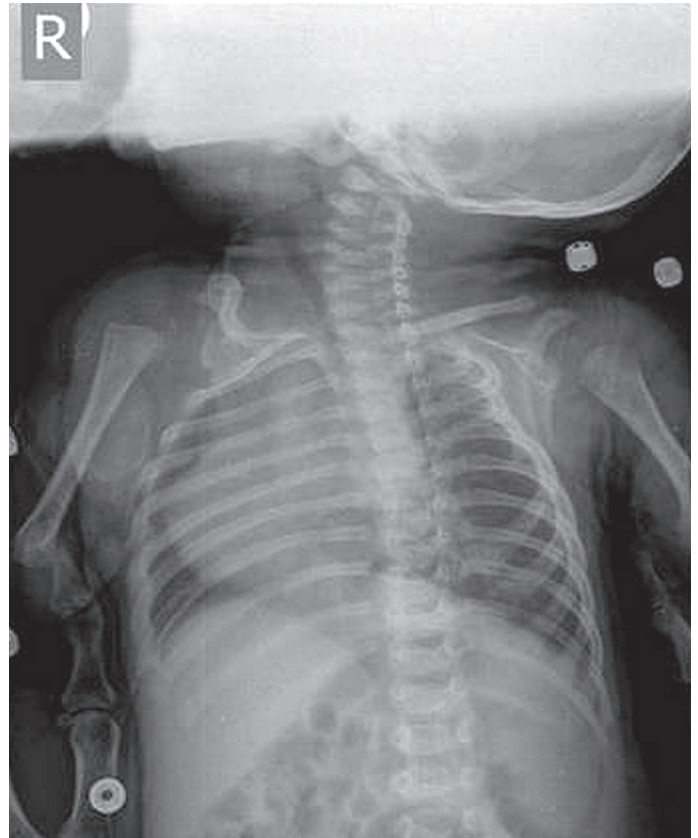


FIGURE 2. Chest X-ray demonstrating dextrocardia and left-sided chest wall deformities.

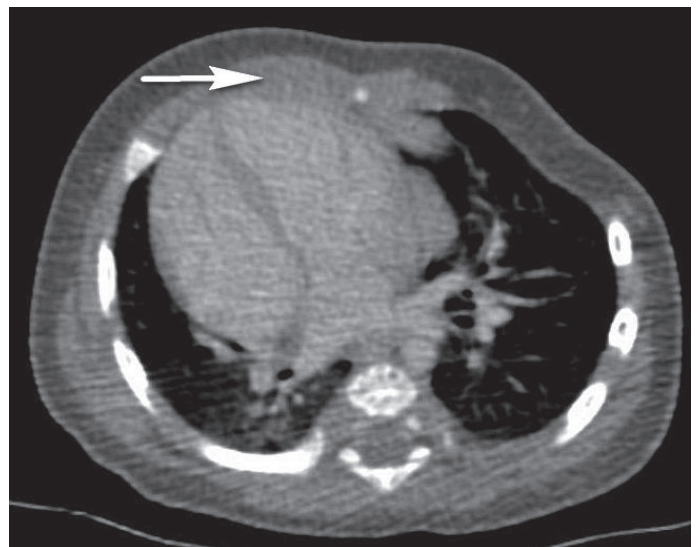


FIGURE 3. Asymmetry of right sided pectoralis muscles (white arrow) with absence of left pectoralis major and minor.

medical history, his mother had primary antiphospholipid syndrome (APS) and she had four intrauterine ex-
tuses. There was no family history of similar complaints or

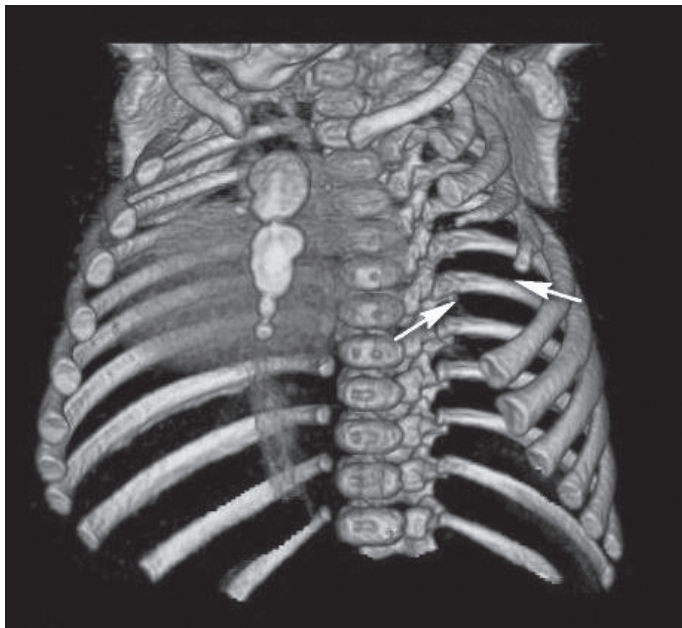


FIGURE 4. 3D volume rendered CT showing hypoplasia of left anterior ribs (white arrow).

findings. Based on these clinical and radiological findings, the diagnosis of Poland's syndrome was established.

DISCUSSION

The pathophysiological basis of the Poland's syndrome is not clear. Subclavian arterial blood supply is suggested to be arrested in early embryonic period leading to hypoplasia of the pectoralis muscles and costal cartilages [6]. Other manifestations of the Poland's syndrome may be caused by involvement of different branches of subclavian artery.

Isolated dextrocardia and associated left-sided anomalies are rarely seen in Poland's syndrome, as far as we know; only 56 such cases have been reported in the literature. Isolated dextrocardia is almost always associated with left-sided Poland's syndrome and left-sided partial rib agenesis [2, 7]. The cause of the occurrence of dextrocardia in Poland's syndrome is not well-known. It may hypothesized that dextrocardia is present at the beginning and resulting transposition of the vascular structures leads to decreased blood supply causing characteristic features of Poland's syndrome. In another hypothesis, dextrocardia may be caused by displacement of the heart to right hemithorax to increase the volume of left side affected by chest wall deformities [5, 7].

Although the patient had facial asymmetry and dextrocardia, hemifacial microsomia was excluded from differential diagnosis because of the absence of other pathognomonic features of hemifacial microsomia such as cranifacial (underdevelopment of external ear, middle ear, mandible, muscles and soft tissue of face) or vertebral anomalies [8].

Hemangioma was only once reported in association with right-sided Poland's syndrome [9]. It is not known whether hemangioma is a component of Poland's syndrome or just an incidental finding.

Maternal thrombophilia associated with primary APS and the presence of antiphospholipid antibodies and thrombophilia in the mother may be responsible for the development of Poland's syndrome by the way of thrombo-occlusive vascular manifestations in the fetus [10]. However, further studies are needed to demonstrate the possible pathophysiological link between APS and Poland's syndrome.

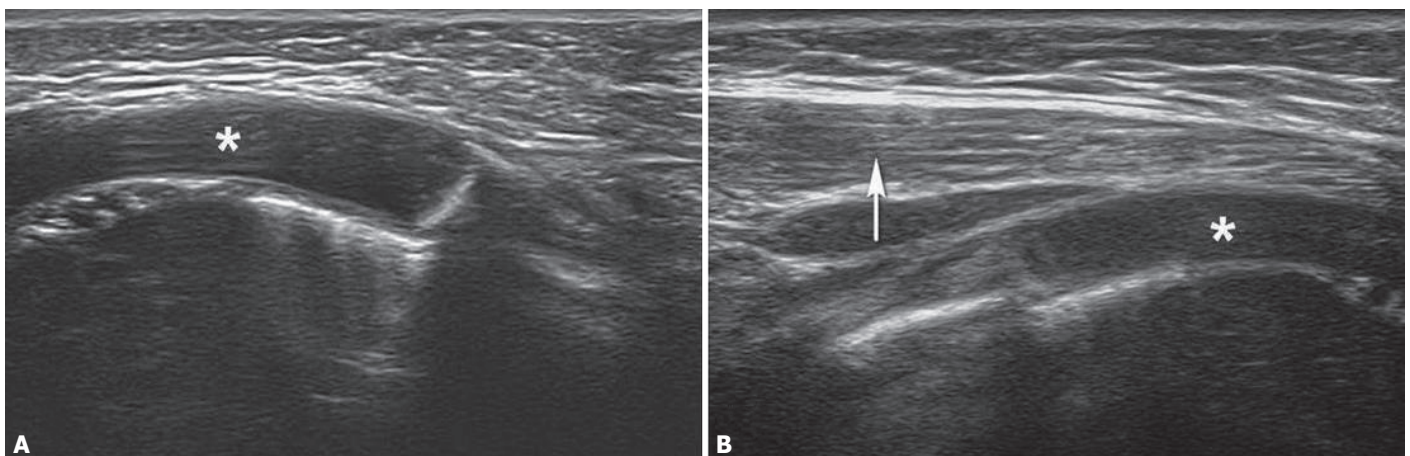


FIGURE 5. Hypoplasia of the left pectoralis muscles compared with right side (white arrow) on ultrasonography.

In conclusion; we have presented a rare case of left-sided Poland's syndrome associated with dextrocardia and hemangioma. Laterality of the Poland's syndrome may give clues about the possible accompanying cardiac malformations.

Informed Consent: Written informed consent was obtained from the parents of the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflict of Interest: No conflict of interest was declared by the authors.

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