

Left-sided Poland's syndrome associated with dextrocardia

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ABSTRACT

Poland's syndrome is a rare congenital anomaly accompanied by the absence of the pectoralis major, and the ipsilateral upper-limb and chest wall deformities. Hypoplasia of the breast, agenesis of the ipsilateral rib cartilage, athelia, and ipsilateral developmental finger anomalies such as syndactyly can also be seen. In the literature, only 56 cases of 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 and partial rib agenesis, or hypoplasia and brachysyndactyly [1]. Hypoplasia or aplasia of the serratus anterior muscle, latissimus dorsi muscle, and external oblique muscle has also been described. Poland's syndrome mostly involves the right side of the chest wall. Anomalies associated with 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]. It is more common

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

CASE REPORT

A 2-month-old male baby was admitted with congenital hemangioma on the tip of the nose (Fig. 1). He was the sixth child from a consanguineous marriage. His weight was 4.9 kg (25th–50th percentile), his height was 57 cm (25th–50th percentile), and occipital circumference was 39 cm (50th–75th percentile). The 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. The chest X-ray showed dextrocardia and left-sided chest wall defor-

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

mities were detected (Fig. 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. An axial chest CT confirmed dextrocardia and demonstrated that right heart chambers were positioned posteriorly, left heart chambers anteriorly, and descending aorta was located on the left with normally connected vascular structures. There was asymmetry of the right-sided pectoralis muscles (white arrow) (Fig. 3). Hypoplasia of left-sided anterior ribs was demonstrated with a 3D-volume rendered CT scan (Fig. 4). Hypoplasia of the left pectoralis muscle was confirmed by ultrasonography (Fig. 5). The renal and testes ultrasonographic examination excluded other 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. The 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 the absence of left pectoralis major and minor.

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



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

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

DISCUSSION

The pathophysiological basis of Poland's syndrome is unclear. The subclavian arterial blood supply is suggested to be arrested in an 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 the involvement of different branches of subclavian artery.

Isolated dextrocardia and associated left-sided anomalies are rarely seen in Poland's syndrome, to the best of our knowledge, and 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 dextrocardia in Poland's syndrome is not well known. It may be hypothesized that dextrocardia is present at the beginning, and the resulting transposition of the vascular structures leads to decreased blood supply causing characteristic features of Poland's syndrome. According to another hypothesis, dextrocardia may be caused by displacement of the heart to right hemithorax to increase the volume of the 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 craniofacial (underdevelopment of the external ear, middle ear, mandible, muscles, and soft tissue of the face) or vertebral anomalies [8].

Hemangioma was reported only once 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 via 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.

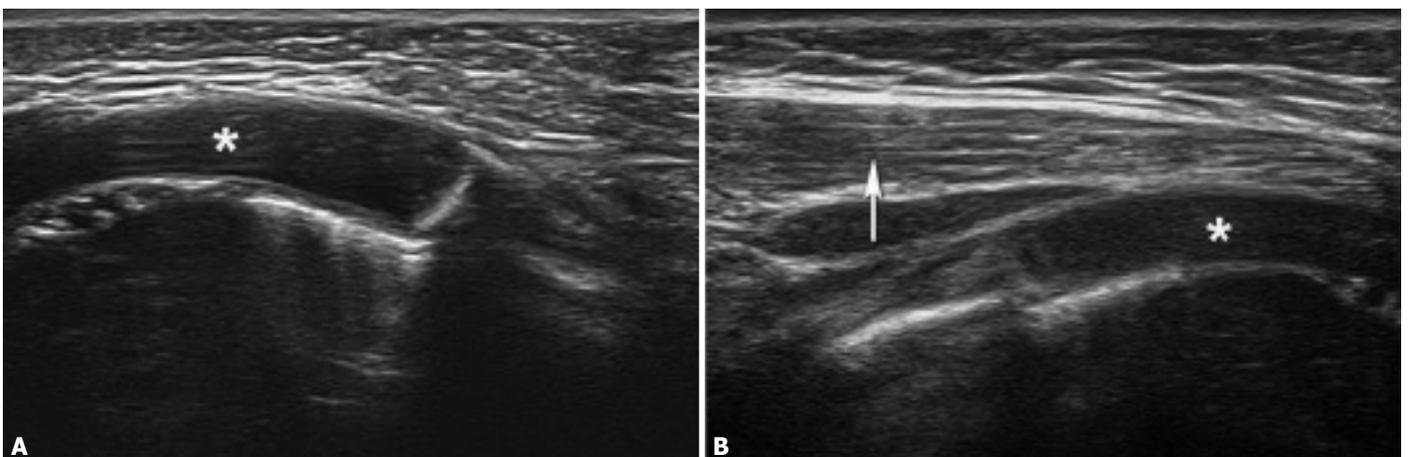


FIGURE 5. Hypoplasia of left pectoralis muscles compared with right side (white arrow) on ultrasonography.

In conclusion, we presented a rare case of left-sided Poland's syndrome associated with dextrocardia and hemangioma. The laterality of 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.

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