CASE REPORT

Poland syndrome involving the left hemithorax with dextrocardia and herniation of the spleen

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SUMMARY

Poland syndrome is characterised by unilateral absence of the large pectoral muscle, ipsilateral symbrachydactyly and occasionally other malformations of the anterior chest wall and breast. The condition is more frequent among men and usually occurs on the right hemithorax in the unilateral form. This case is unique because we believe it is a rare case of Poland syndrome involving the left hemithorax along with dextrocardia and herniation of the spleen from the left subcostal region.

BACKGROUND

Alfred Poland, in 1841, first described a new syndrome characterised by complete unilateral absence of the pectoralis major, serratus anterior and external abdominal oblique muscles associated with ipsilateral thoracic and upper limb defects.¹ The spectrum of thoracic defects in Poland syndrome includes a wide range of anomalies from subtle hypoplasia to aplasia of the breasts and ribs, chest wall depression, sternal anomalies, absence of axillary hair and limited subcutaneous fat. The classic hand deformity includes syndactyly and variable degree of brachydactyly and severe hypoplasia or aplasia of the middle phalanges. Our case is unique because we believe it is a rare case of Poland syndrome involving the left hemithorax along with dextrocardia and herniation of the spleen from the left subcostal region.

CASE PRESENTATION

Our case is a 1-month-old male child who presented to our paediatric outpatient department with abnormal chest wall configuration. The patient was born after an uneventful first pregnancy to a healthy and non-consanguineous couple. Family history of abnormalities of limbs or thorax of both parents was negative. Physical examination showed a 4×3 cm defect in the left subcostal region along with herniation of the spleen (figure 1A) and asymmetrical hypoplasia of the left hemithorax (figure 1B) with a hypoplastic left nipple along with polysyndactyly on upper limbs (figure 2A) and lower limbs (figure 2B). The right upper extremity was normally developed. The lower extremities and genitalia were unremarkable. The patient appeared clinically normocephalic without any dysmorphic features. The neurological examination was normal, with good sensation and symmetric movement of all extremities, spontaneously and to stimulation. Primitive reflexes were intact. The pupils were reactive to light and accommodation with normal motion aftereffects. On examination of the abdomen, a 4×3 cm defect was seen in the left

Figure 1 Physical examination of the abdomen and chest showing a 4×3 cm defect (black arrow heads) seen in the left subcostal region along with herniation of the spleen (black asterisk) (A) and asymmetrical hypoplasia of the left hemithorax (black thick arrow) (B).

subcostal region along with herniation of the spleen (figure 2A,B). Apical impulse was located on the right hemithorax. There was no history of respiratory distress.

INVESTIGATIONS

The child underwent radiographic examinations, including chest X-ray and CT scans of the chest and abdomen which identified several anatomical abnormalities. Chest X-ray (figure 3A) and subsequent three-dimensional thoracic CT-scan (figure 3B) showed partial agenesis of the left fourth, fifth and sixth rib along with eventration of the diaphragm, dextrocardia and herniation of the spleen through the defect. No lung parenchyma deformities were seen.

TREATMENT

Since the child is asymptomatic and thriving well, a conservative approach is being considered with close follow-up. The parents were advised to avoid trauma to the site of herniation of the spleen.

Figure 2 Physical examination of the upper and lower limbs showing polysyndactyly of the right upper limb (black asterisk: polydactyly finger attached to the little finger, syndactyly of the middle and ring finger, thumb not shown in figure) (A) and polysyndactyly of great toes of the lower limbs (black asterisk: polysyndactyly of the right great toe; black thick arrow: polysyndactyly of the left great toe) (B).

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Figure 3 Chest X-ray and CT scan of the chest and abdomen showing asymmetrical hypoplasia of the left hemithorax (white thick arrow), dextrocardia (white thin arrow) (A) and herniation of the spleen through the defect (white asterisk) (B).

OUTCOME AND FOLLOW-UP

The child is under close follow-up and is asymptomatic with normal milestones of development at 6 months of age. No local complications at the site of herniation of the spleen were found. The child is planned for anatomical or mesh repair of the splenic hernia defect. The authors are not in favour of splenectomy as the child is very small and splenectomy in this age group has its own complications.

DISCUSSION

The incidence of Poland syndrome is 1 out of 30 000 people, with a male predominance. Most cases have been described as sporadic, although some familial cases have been reported. The right side is affected in 60–75% of patients,² and cases of bilateral absence of the pectoralis major muscle have been reported in the literature.³ Although the exact cause of Poland syndrome is unclear, most researchers believe the origin to be vascular. Interruption of the subclavian artery or its branches at the sixth week of gestation is believed to result in the clinical manifestation of the disease. The association of Poland syndrome with dextrocardia has been reported in 36 cases, and the incidence of this concurrent condition is 5.6%–11.5%.

Congenital subcostal spleen herniation without a chest wall defect is a separate entity, generally related to diaphragmatic eventration. Eventration of the diaphragm in association with Poland syndrome and wandering spleen syndrome without herniation have been described in the literature.⁴ Our case is a rare case of Poland syndrome involving the left hemithorax along with dextrocardia, eventration of the diaphragm and herniation of the spleen from the left subcostal region though the association of Poland syndrome with herniated liver is reported in the

literature.^{5–8} Like any other herniated organ, a herniated splenic segment runs the risk of incarceration with subsequent necrosis. In addition, the narrow and long spleen stalk makes the lesion susceptible to injury, carrying the risk of significant bleeding. This case enriched the clinical perception of Poland syndrome and provided us an opportunity to understand the exact cause and underlying mechanisms of various complicating abnormalities associated with the disease.

Learning points

- Poland syndrome is a rare birth defect characterised by underdevelopment or absence of the pectoralis muscle on one side of the body, and usually also ipsilateral symbrachydactyly.
- The condition is benign, and may be asymptomatic depending on the degree of involvement of the upper extremity.
- Poland's syndrome occurs sporadically ('out of the blue') and does not run in families. The risk of recurrence of Poland's syndrome in the family is minute (1%).
- Poland syndrome with herniated liver is reported in the literature and its association with subcostal herniation of the spleen is the first seen in our case.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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