

Poland Anomaly with Dextrocardia: A Case Report

Pages with reference to book, From 181 To 182

Tabish Hazir, Munazza Saleem Malik (The Children's Hospital, Pakistan Institute of Medical Sciences, Islamabad.)

Introduction

Poland anomaly was first reported in Guys Hospital report, 1841¹. Since then many cases have been described. Among the upper limb developmental defects, Poland anomaly has a prevalence of 1 in 20,000. According to another report 10% of all cases of upper limb syndactyly have Poland anomaly² also. Poland anomaly is a sporadic developmental disorder involving the upper limb. Primary defect seems to be in the development of the proximal subclavian artery, with early deficit of blood flow to the distal limb and the pectoral region, yielding partial loss of tissue in those regions³. There is a male preponderance with 70% involving the right side. The clinical spectrum is variable depending upon the extent of expression. Upper limb defect is distal agenesis of varying severity with syndactyly being the commonest presentation. Thoracic deformities include hypoplasia and absence of subcutaneous tissue of anterior chest on the affected side; absence of hypoplasia, with displacement of nipple and breast; absence of sternocostal part of pectoralis major muscle; absence of pectoralis minor and costal cartilage abnormalities⁴.

Case Report

One hour old female baby, born full term with birth weight of 2.7Kg, was shifted to neonatal intensive care unit of Children's Hospital on account of multiple congenital abnormalities. On examination, left pectoralis major muscle was absent which resulted in the obvious depression of the chest wall on the left side. There was displacement of right nipple. There was severe distal hypoplasia of the left upper limb with rudimentary hand consisting only of skin tags attached to the forearm. On auscultation, heart sounds were heard on the right side of the chest. Rest of the physical examination was unremarkable. X-ray chest showed dextrocardia along with malformed and mal-aligned left upper ribs which were normal in number. Echocardiography confirmed dextrocardia without associated cardiac abnormalities. Ultrasound of abdomen was normal with no renal abnormality.

Comments

Since the first case report in 1841, many cases have been described with varying features. Takayama and Ireland et al reviewed 43 cases of Poland anomaly⁵. Mace, Kaplan et al reported seven cases⁶. In these case reports, limb and thoracic cage defects are well recognized and virtually constant features are anomalies of the nipple, subcutaneous tissue and pectoralis major and minor. Upper limb defects with varying degree of severity include syndactyly, brachydactyly, oligodactyly and occasionally more severe reduction deficiency of the upper limb. In addition to these consistent abnormalities, there are certain less common associated abnormalities which are present in some patients of Poland anomaly. Occasional associated abnormalities include renal abnormalities⁷, hemivertebrae, scoliosis and Moebius syndrome exhibiting paralysis of the sixth and seventh cranial nerves⁸. None of these case reports mention the association of Poland anomaly with dextrocardia which was present in our patient.

References

1. Poland, A. Deficiency of pectoral muscles. *Guy's Hospital Rep.* 1841. In Kenneth Lyons Jones (Ed.) *Smith's recognizable patterns of human malformations*, 4th ed, Philadelphia, W.B. Saunders Co., 1988, pp. 260-61.
2. Claenckson. P. Poland syndactyly. *Guy's Hosp. Rep.*. 1962. In Jones K.L. (Ed.) *Smith's recognizable pattern of Human malformations*. 4th Ed. Philadelphia, W.B. Saunders Co.. 1988. pp.260-61.
3. Bouvet, J., Maroteaux, P. and Briard-Guillemot, M. Poland's syndrome: Clinical and genetic studies - physiopathology. *Nouv. Presse. Med.*, 1976;5: 185-7.
4. Goodman, R.M. and Gorlin, R.J. Poland anomaly. *The malformed infant and child*. New York. Oxford University Press, 1983, pp. 74-75.
5. Ireland, D.C.R., Takayama, N., Flat, A.E. Poland syndrome. A review of 43 cases. *J. Bone Joint Surg.*, 1976;58 A:52-58
6. Mace, J.W., Kaplan, J.M., Shanenbergen, I.E. et al. Poland syndrome. Report of seven cases and review of literature. *Clin, Pcdiatr.*. 1972;1 1:98-102.
7. Hoover, G.H., Flatt, A.E. and Weiss, MW. The hand and Apert syndrome. *J. Bone Joint Surg.*, 1970;52 A:878-892.
8. Goodman, R.M. and Gorlin, R.J. Poland anomaly. *The malformed infant and child*. New York. Oxford University Press, 1983, pp. 74-75.