International Surgery Journal
Reshma S et al. Int Surg J. 2017 Oct;4(10):3526-3528
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20174530

Case Report

Poland's syndrome: a rare case report

Reshma S.*, Vijai R., Chakarvarthy N.

Department of General Surgery, Saveetha Medical College and Hospital, Thandalam, Chennai, Tamil Nadu, India

Received: 26 July 2017
Accepted: 20 August 2017

***Correspondence:**
Dr. Reshma S.,
E-mail: surgeonreshma@gmail.com

ABSTRACT

Poland's syndrome is a rare congenital condition. It is classically characterized by absence of unilateral chest wall muscles and sometimes ipsilateral symbrachydactyly (abnormally short and webbed fingers). The condition typically presents with unilateral absence of the sternal or breast bone portion of the pectoralis major muscle which may or may not be associated with the absence of nearby musculoskeletal structures. We report a 25-year-old male patient with typical features of Poland's syndrome. To the best of our knowledge, this is the first documented case of a patient with Poland's syndrome reported from Chennai.

Keywords: Hypoplasia, Poland's syndrome, Pectoralis major

INTRODUCTION

The Poland's anomaly was first described in 1841 by Sir Alfred Poland as a syndrome presenting with absence or underdevelopment of pectoralis major muscle, associated in some cases with a hypoplasia of the breast, an agenesis of 2,3,4 and 5 ipsilateral costal cartilage, an athelia, and an ipsilateral webbing of the fingers (cutaneous syndactyly).[1,2]

Therefore, the Poland's syndrome may occur with different gravity. Currently, it is assumed that Poland's syndrome is characterized by a missing sternocostal bundle of the pectoralis major muscle.[3] Its incidence is difficult to determine, but current estimates range between 1:7000 and 1:100000 births, with higher frequency among males (ratio, 2:1-3:1). In 75% of the cases, it is located on the right hemithorax in the unilateral form.[1,3-6] Affected individuals may have variable associated features, such as underdevelopment or absence of one nipple including the areola and/or patchy absence of hair in the axilla.[4] In females, there may be underdevelopment or aplasia of one breast and underlying (subcutaneous) tissues.[7]

In some cases, associated skeletal abnormalities may also be present, such as underdevelopment or absence of upper ribs; elevation of the shoulder blade (Sprengel deformity); and/or shortening of the arm, with underdevelopment of the forearm bones (i.e., ulna and radius).[1,4] We present the first documented case from Chennai. This paper is aimed at bringing awareness to health professional in the region of this rare congenital condition.

CASE REPORT

A 25-year-old male presented to surgery OPD with a flattening of his right anterior chest wall since birth. There was no familial history of similar disorder. Examination revealed, normal growth parameters and good mental status. His chest was asymmetric, with hypoplasia of the right side. He had a shapeless right shoulder, flattening of the right chest wall and absence of the right axillary fold. The pectoralis major muscle (Figure 1) was absent but the pectoralis minor as well as the serratus anterior muscle were present. The movements of the right shoulder were possible with the muscle power at abduction estimated at 4/5. He had

normal heart sounds, respiration and breath pattern. The limbs were normal and symmetric, and ipsilateral fingers were found to be normal. Radiological examination of the chest showed no abnormalities of the ribs or heart. X-ray of bones of the ipsilateral upper limb was normal. Based on those physical findings, a diagnosis of Poland's syndrome was made. No surgical treatment was offered. The family was counseled and the patient followed up on outpatient basis. During the last two years of follow up, patient is doing well and found to be normal.

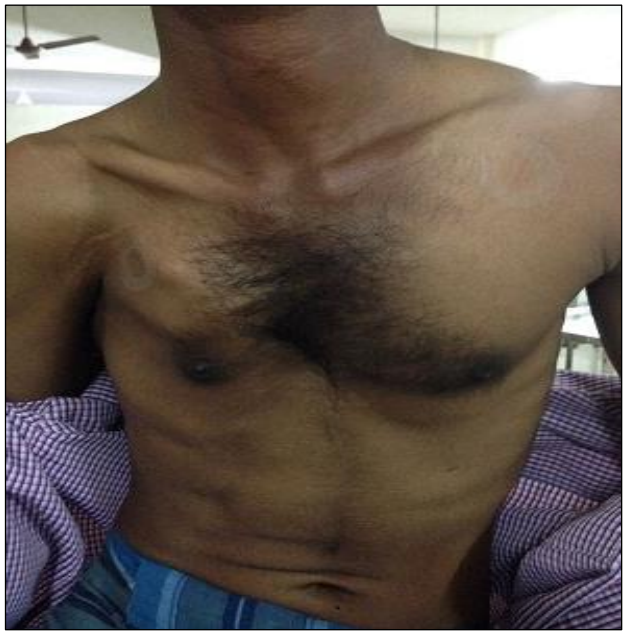

Figure 1: Absence of pectoralis major muscle.

DISCUSSION

The case of Poland's syndrome we present, is the first described in Chennai and is of the pure presentation as it consists only on the unilateral aplasia of the pectoralis major muscle without any other associated defects.[1,3] The exact etiology of the Poland's syndrome is unknown. It is assumed that the aplasia of the pectoralis muscles and associated chest defects, as the athelia, aplasia of costal cartilages, are consequences of an interruption of early embryonic blood supply of sub-clavicular artery branches.[5,6] A combination of the blockage of various branches could lead to different manifestations of the Poland's syndrome. It is known that thoracic wall is supplied by medial thoracic branches, intercostals artery, and the thoracic artery from axillary artery, the thoraco-acromial artery and the lateral thoracic artery. All these branches come from the subscapular artery or axillary artery. The interruption of the blood supply is caused by thrombus or embolus, which prevent the blood to reach the developing tissue. Another cause of blood supply interruption is the mis-development of vessels. However, there have been case reports of Poland's syndrome associated with unusual defects, which cannot be explained on the basis of compromised blood supply alone. On the other hand, Ferraro and colleagues described an unusual presentation of the Poland's anomaly without any vascular alteration, raising the question as to the true pathogenesis of the Poland's syndrome.[3] Geneticists currently hold the view that Poland syndrome is rarely inherited and generally is a sporadic event. There are rare instances where more than one individual has been identified with Poland's syndrome either in the immediate, or extended family.[6,8-12] Therefore, some authors believe that an inherited abnormal vasculature formation may be the central underlying mechanism for this condition. Several reconstructive procedures are available to correct the functional and structural deformities associated with this syndrome. As for the chest deformity, customized silicone prosthesis is simply and safely used. Transposition of the latissimus dorsi muscle for soft-tissue reconstruction has been used by many authors with satisfactory esthetic and functional results.[13]

CONCLUSION

This case report adds to the knowledge of health professionals in the region about this rare congenital condition.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Fokin AA, Robicsek F. Poland syndrome revisited. Ann Thorac Surg. 2002;74:903-18.
2. Lagbo JN. Poland syndrome: report of a variant. J Natl Med Assoc. 2006;98:97-9.
3. Ferraro GA, Perrotta A, Rossano F, D'Andrea F. Poland syndrome: description of atypical variant. Aesth Plast Surg. 2005;29:32-3.
4. Urschell HC. Poland syndrome. Chest Surg Clin. North Am. 2000;10:393-403.
5. Bavinch JNB, Weaver DD. Subclavian artery disruption sequence: hypothesis of vascular etiology for Poland, Klippel-Feil and Mobius anomalies. Am J Med Genet. 1986;23:903-18.
6. Cobben JM, Robinson PH, van Essen AJ, van der Wiel HL, ten Kate LP. Poland anomaly in mother and daughter. Am J Med Genet. 1989;33:519-521.
7. Azner JMP, Urbano J, Laborda EG, Moreno PQ, Vergara LF. Breast and pectoralis muscle hypoplasia: a middle degree of Poland syndrome. Acta Radiologica. 1996;37:759-62.
8. Czeizel A, Vitéz M, Lenz W. Birth prevalence of Poland sequence and proportion of its familial cases. Am J Med Genet. 1990;36:524.
9. Hoyme HE, Der Kaloustian VM, Hogg H, Entin MA, Guttmacher AE. Possible common pathogenetic mechanisms for Poland sequence and Adams-Oliver syndrome: an additional clinical observation. Am J Med Genet. 1992;42:398-9.

10. David TJ. Familial Poland anomaly. J Med Genet. 1982;19:293-6.
11. Lowry RB, Bouvet JP. Familial Poland anomaly. J Med Genet. 1983;20:152-4.
12. Fraser FC, Ronen GM, O'Leary E. Pectoralis major defect and Poland sequence in second cousins: extension of the Poland sequence spectrum. Am J Med Genet. 1989;33:468-70.
13. Freitas RS, Tolazzi ARD, Martins VDM, Knop BA, Graf RM, e Cruz GAO. Poland syndrome: different clinical presentations and surgical reconstructions in 18 cases. Aesth Plast Surg. 2007;31:140-6.

Cite this article as: Reshma S, Vijai R, Chakarvarthy N. Poland's syndrome: a rare case report. Int Surg J 2017;4:3526-8.