

Case Report

A rare case of Poland: Mobeius syndrome in an infant

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Received: 31 May 2019

Revised: 04 July 2019

Accepted: 09 July 2019

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ABSTRACT

Mobius syndrome is a rare condition of unclear origin, characterized by a unilateral or bilateral congenital facial weakness with impairment of ocular abduction, which is frequently associated with limb anomalies. Poland Syndrome is a rare condition that is evident at birth (congenital). Associated features may be extremely variable from case to case. However, it is classically characterized by absence (aplasia) of chest wall muscles on one side of the body (unilateral) and abnormally short, webbed fingers (sybrachydactyly) of the hand on the same side (ipsilateral). In those with the condition, there is typically unilateral absence of the pectoralis minor and the sternal or breastbone portion of the pectoralis major. In females, there may be underdevelopment or absence (aplasia) of one breast and underlying (subcutaneous) tissues. 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). Other associated abnormalities may include dextrocardia, diaphragmatic hernia and renal anomalies etc. Poland Syndrome affects males more commonly than females and most frequently involves the right side of the body. The exact cause of the condition is unknown. The combination of Poland-Mobius syndrome is rare, with an estimated prevalence 1:500 000.

Keywords: Mobius syndrome, Pectoralis, Poland syndrome

INTRODUCTION

Poland Syndrome is a rare congenital disorder. Associated features may be variable from case to case. However, it is classically characterized by absence (aplasia) of chest wall muscles on one side of the body (unilateral) and abnormally short, webbed fingers of the hand on the same side. Poland syndrome is not an inheritable condition nor one that arises from events during pregnancy, which was established in a detailed study of twins affected with Poland syndrome using DNA evidence to establish that they were identical.¹ The non-genetic and non-teratogenic etiology suggests that Poland syndrome is entirely sporadic and combinations of Poland and Mobius anomalies are rarely described in the

literatures with an estimated prevalence of 1:500000.^{2,3,4} Some authors believe this association is an independent syndrome and Disruption of the subclavian artery occurs around week 6 of gestation may be related to Moebius–Poland syndrome.

CASE REPORT

This case involves a 4 months old male baby brought by the mother to the OPD with the complaints of asymmetric chest and right chest wall depression. There were no breathing or cardiac complaints on review of systems, but the child had been admitted for 7 days last month and was diagnosed with pneumonia. The child was born by normal vaginal delivery, term gestation with a birth

weight of 3 kg and cried immediately after birth with uneventful postnatal phase. Antenatal period was also uneventful with no history of any drug intake, hypertension or diabetes mellitus. The child was 1st child of the parents and was developmentally normal with growth parameters appropriate for his age. Physical examination showed chest asymmetry with right anterior chest wall depression and flattening of the right pectoral region (Figure 1).



Figure 1: Infant with hypoplastic pectoralis major showing right sided flattening and chest wall recessions.

The child had right lateral chest wall recessions on breathing, but the air entry was bilaterally equal and there were no added sounds. Abduction of the shoulders showed absence of the sternocostal head of pectoralis major. Hand examination did not show any signs of ipsilateral digital abnormality. Also, the child was not able to abduct his right eye hence it was a combination of Poland and Moebius syndrome which was an exceptional finding.

DISCUSSION

Moebius syndrome consists of congenital complete or partial facial nerve paralysis with or without paralysis of other cranial nerves. The most common cranial nerve involved is the 6th cranial nerve (abducent). The oculomotor and trochlear can also be involved. It is also known as congenital facial diplegia, nuclear agenesis, congenital nuclear hypoplasia, congenital oculofacial paralysis and congenital abducens-facial paralysis.⁵ According to the pathologic changes, Moebius syndrome is classified into four groups.⁶ Group I is characterized by simple hypoplasia or atrophy of cranial nerve nuclei, presumably as a result of embryonic maldevelopment. Group II results from primary lesions in the peripheral portion of the cranial nerves. Group III is due to focal necrosis in the brain stem nuclei. Group IV consists of patients without lesions in the central nervous system or cranial nerves but showed features of primary myopathy.

The Moebius syndrome represents a distinctive malformation syndrome, but its etiology remains undetermined. Numerous theories exist concerning the primary underlying pathogenesis. Intrauterine vascular etiology involves disruption of the flow in the basilar artery or premature regression of the primitive trigeminal arteries.⁷ Congenital inherited hypoplasia or agenesis of the CN nuclei, and infectious factors have been also proposed as important factors. Poland syndrome was described in 1841 by Alfred Poland as unilateral absence of pectoralis major muscle and ipsilateral dermal syndactyly of the hand.⁸ Other anomalies associated with this syndrome include hypoplasia of the forearm, hypoplasia of the breast, agenesis of the nipple, rib cage deformities, bilateral epicanthus and talipes equinovarus.⁵ Boys are affected more than girls. The reported frequency is 1:20000.⁵ Ten percent of all cases of hand syndactyly may have the Poland sequence. The syndactyly is usually in the right hand as in this patient. Poland syndrome is not an inheritable condition nor one that arises from events during pregnancy, which was established in a detailed study of twins affected with Poland syndrome using DNA evidence to establish that they were identical.¹ Furthermore, since the both of the embryos, fetuses and newborns have been exposed to identical physical and chemical conditions, there were no environmental teratogenic sources implicated. This non-genetic and non-teratogenic etiology suggests that Poland syndrome is entirely sporadic. combinations of Poland and Moebius anomalies are rarely described in the literatures with an estimated prevalence of 1:500 000.^{2,4} Some authors believe this association is an independent syndrome.

Disruption of the subclavian artery occurs around week 6 of gestation and is related to Moebius–Poland syndrome; this same phenomenon is related to transversal limb defects and arthrogryposis.⁸ The disruption phenomenon can be secondary to a blood flow interruption secondary to an arterial spasm during the sensitive embryonic phase.⁹

The abnormalities in the vascular structure could be secondary to teratogenic effects. Teratogens can act directly by reducing blood flow or blood vessel development, changing anatomy and/or structure.^{10,11} The vascular abnormalities of the right subclavian artery observed in Poland syndrome could be related to a vascular disruption caused by misoprostol during a critical period.¹²

CONCLUSION

It is a rare inherited disorder with aplasia of pectoralis major muscle on either side with or without associated disorders. 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 and does not run in

families. The risk of recurrence of Poland's syndrome in the family is 1%.

A team approach is required for management of patient with Poland syndrome. 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

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Gupta A. A rare case of Poland: Mobeius syndrome in an infant. *Int J Contemp Pediatr* 2019;6:2206-8.