IJPMR 2008 October; 19 (2):61-63 **Case Report**

A Rare Case of Poland Syndrome with Absence of Ipsilateral Nasolacrimal Duct

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Introduction

First described in a cadaver examined by A. Poland at Guy's Hospital in 1841, "this combined anomaly of webbed fingers(cutaneous syndactyly) of the hand on the same side and the absence or underdevelopment of the sterno-costal portion of ipsilateral pectoralis major muscle is known as Poland's Anomaly, Poland's Syndactyly, or Poland's Syndrome.[1] It is a rare condition present at birth (congenital). For people born with Poland's Syndrome, the breastbone portion (sternal) of the pectoralis is also missing.

Case Report

A 13-year-old boy presented with complaints of weakness on the right side along with maldevelopment of right hand since birth and epiphora since birth. H/O antenatal bleeding was there and some drug was given was a Dr. at village and her mother is unable to recall that drug. Otherwise family history, past history are not suggestive .Other siblings (4 in number) are normal.

On examination, child was thin built, weighing 35 kg and height was 129 cm. Arm span was 122 cm and arm span (measured from mid-point of sternum) was 65 cm on left side and 57 cm on right side. Others vital were within normal limit.

There was facial asymmetry on right side. Nasal bridge was flattened. There was absent nasolabialfold, wrinkle free forehead, incomplete closure of eye on right side. There was absent right sided mammary gland and right pectoralis major. There was symbrachydactyly of right hand. Sternum was bifid. Right sided 5th and 6th ribs were absent. There was no scoliosis. Sprengal's deformity of right shoulder was there. First web space- enlarged between bilateral toes.On CVS, P/A, Genitourinary examination no abnormality was detected clinically. Also there was platynychia of both lower limbs toes. One ectopic tooth was present behind upper central incisor teeth. On neurological examination, there was decrease in strength of all muscles of right upper limb and right sided facial nerve palsy. Although patient's right hand function was impaired but he was able to perform ADL with left hand.

On ocular examination, B/L Bitot's spots were detected with refractive error of

-1.0 d. Slit lamp biomicroscopy revealed that upper and lower puncta of right eye were absent.

Patient and his parents were educated about the disease and progressive resistive strengthening exercises of the right shoulder girdle muscles and facial muscle strengthening exercises and facial massage were prescribed. ADL training was given to the patient. Glasses were prescribed for refracive error.

Discussion

Poland Syndrome is a type of uncommon sporadic congenital syndactyly leading to the absence of the Pectoralis Major muscle and often linked to limb deficiencies.[2] Poland's syndrome, also known as Poland's sequence or Poland's anomaly, consists of many distinctive features. The most prominent, among them are ipsilateral hand anomalies chiefly in the form of syndactyly along with absence of pectorals. Poland's syndrome was first described by Alfred Poland in 1841[3] includes partial or complete absence of pectoralis along with ipsilateral hand anomalies, ranging from mild defects to severe bony abnormalities.[4] The incidence ranges from

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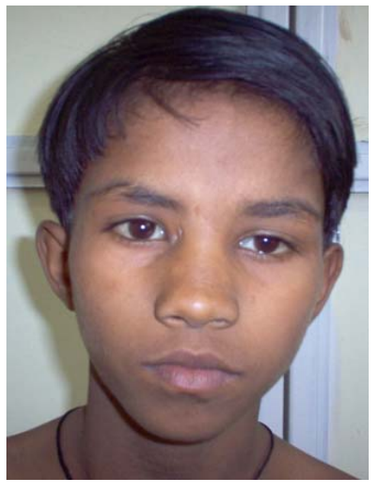

Figure 1: Facial asymmetry on right side

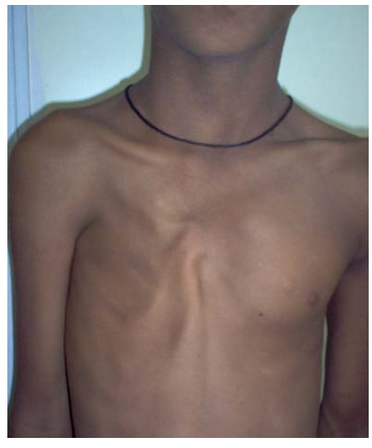

Figure 2: Absence of nipple and right pectoralis major with bifid sternum.Absent right 5-6th rib

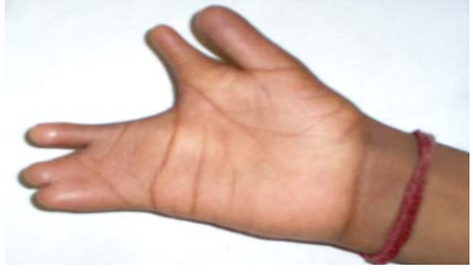

Figure 3: Symbrachydactyly of right hand

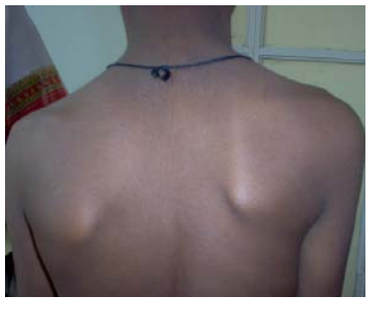

Figure 4: Sprengal's deformity of right shoulder

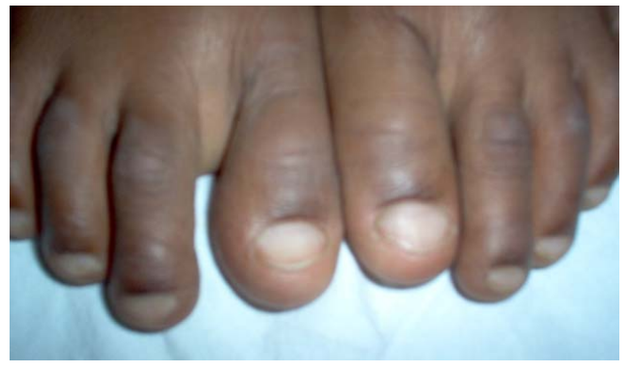

Figure 5: Platynychia

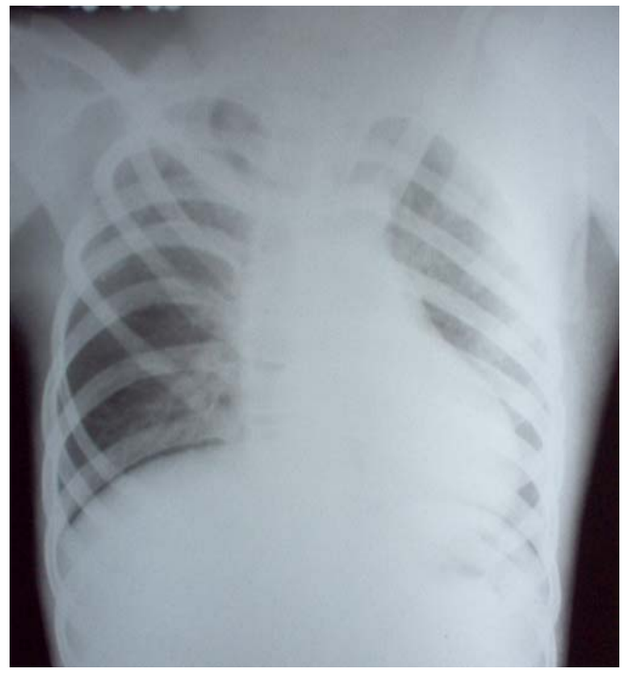

Figure 7: Skiagram of chest-PA view showing absence of right 5th & 6th rib

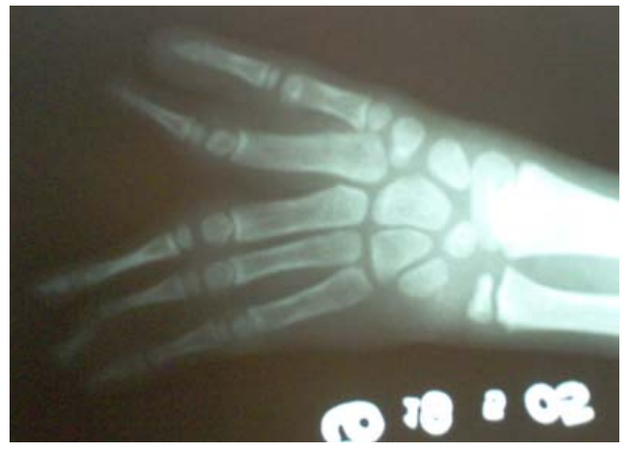

Figure 8: Radiograph of right hand showing hypoplasia of 1st metacarpal and phalanges

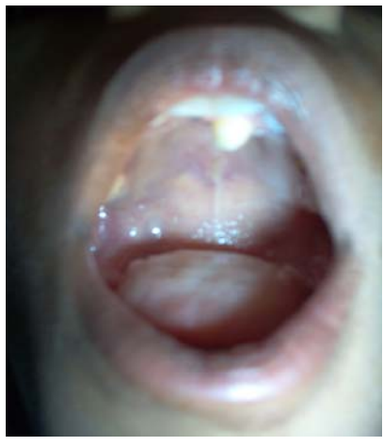

Figure 6: Ectopic tooth

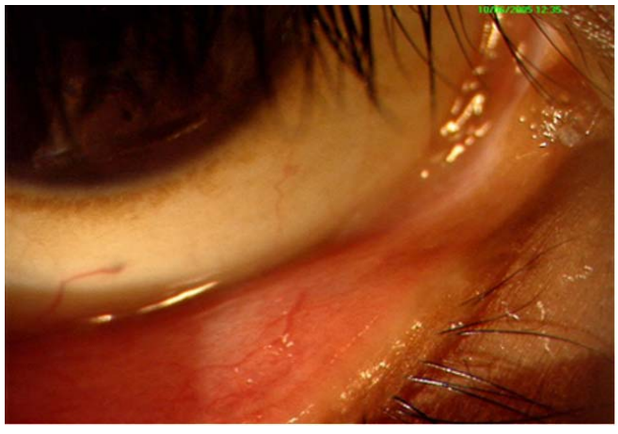

Figure 9: Absent lower puncta in right eye

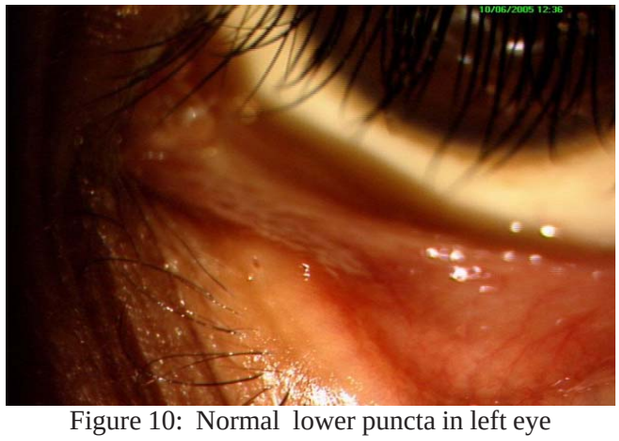

Figure 10: Normal lower puncta in left eye

1:20000 to 1:50000 as reported by different authors. Males are more often affected than females by nearly 2:1. The effects are seen on the right side most often by nearly 5:3.

Most researchers currently find that there is no familial cause, but there are some who link this Syndrome with several others, and hypothesise a circulatory defect in the subclavian artery during weeks 5 to 7 of pregnancy, and connect this with the action of a particular autosomal dominant gene.[5] A combination of the blockade of various branches could lead to Poland syndrome along with its variants. These vascular disruptions could also lead to its relation with syndromes like Sprengel, Klippel-Fiel and Adams-Oliver syndrome.[6]

Very few cases are familial. Most cases are sporadic, as this one. The inheritance can be autosomal dominant; however variable expressivity and reduced penetrance is usually present. Most case reports describe affliction of a single side. There have been case reports of Poland syndrome associated with unusual defects which cannot be explained on the basis of compromised blood supply alone encompassing a host of abnormalities such as dextrocardia, genitourinary and spinal malformations and malignancies such as leukemia and non-Hodgkin lymphoma. One such case report described by Kabra, et al., reported a myriad of abnormalities, not interrelated in which the contralateral side has also been affected, in addition to involvement of both the feet.[7]

Conclusion

Poland syndrome has broad spectrum of skeletal, ocular, dental and maxillofacial anomalies. We have not come across any available literature about congenital absence of nasolacrimal duct. Multidisciplinary team approach is required for motor skills and co-ordination to gain better control over activities of daily living and patient counselling about this entity is very crucial.

References

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