CASE STUDY

Poland syndrome associated with contralateral gynecomastia

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Introduction

Poland syndrome is an uncommon congenital birth defect that may present with partial or complete absence of the pectoralis muscle. It is commonly accompanied by nipple-areolar complex, breast abnormalities, thinning of subcutaneous tissue, deformities of ribs, axillary and pectoral hair absence, and unilateral hand syndactyly. Some of these anomalies present at puberty. Out of these, the chest may become more apparent. However, severely affected children could have these anomalies in hand or chest from birth.

Case Report

This seventeen-year-old male presented with a complaint of left side breast enlargement for 2 years' duration. At the age of 2 years, parents noticed his chest wall is not symmetrical, so they sought medical advice and were reassured. Since he was 15 years old, they have noticed a rapid enlargement in his left breast. The patient is non-alcoholic, non-smoker with no history of medication, liver disease, renal disease, or any traumatic injury.

On examination the patient is obese, the thyroid is diffusely enlarged. He has right-sided syndactyly affecting right ring and middle fingers. On local examination, the right nipple and areola complex was smaller and was at a superior position. Moreover, there was no nipple discharge on the left side. On palpation, the abdomen was normal. Both testes were palpable in the scrotal sac and they were normal in size and volume.

Laboratory Investigations:

AFP	**– 6.79 micg/dL (<8.5)**
Testosterone	**– 9.20 nmol/L (9.7 - 38.14)**
FBS	**– 67.2 mg/dL**
Total Cholesterol	**– 283 mg/dL**
TG	**– 122.4 g/dL**
HDL	**– 46.4 mg/dL**
LDL	**– 212 mg/dL**
C: H R	**– 6**
Beta HCG	**<1.2 mIU /ml**
FSH	**– 2.06 mIU/ml (0.7-11.1)**
LH	**– 4.76 mIU/ml (0.8-7.6)**
TSH	**– 2.92 IU/ml (0.4 -4.0)**
T4	**– 1.44 ng/dL (0.89-1.76)**

Patient underwent liposuction with nipple preserving gynecomastia correction surgery.

Discussion

Gynecomastia is commonly defined as a benign excessive development of breast tissues in a male. There are four grades of gynecomastia (6);

Classification

The causes for this condition are physiological causes such as puberty, certain medications, liver diseases, starvation, hypogonadism, certain tumours like prolactin-secreting pituitary adenomas, hyperthyroidism and certain syndromes like Klinefelter syndrome (6). Poland syndrome is quite an uncommon condition encountered in surgical practice which is estimated to occur in 1 in 30 000 patients. The right side of the chest is affected twice as often as the left. In this condition, no gender predilection is exhibited (4).

Grade I	Minimal hypertrophy (<250g of breast tissue) without ptosis I A. Primarily glandular I B. Primarily fibrous
Grade II	Moderate hypertrophy (250-500 g of breast tissue) without ptosis II A. Primarily glandular II B. Primarily fibrous
Grade III	Severe hypertrophy (> 500g of breast tissue) with grade I ptosis Glandular or Fibrous
Grade IV	Severe hypertrophy with grade II or III ptosis Glandular or fibrous

Anomaly in vascular developmental sequence during the 6th week of intrauterine life along with hypoplastic of the subclavian artery is believed to be the aetiology which causes musculoskeletal malformations (4).

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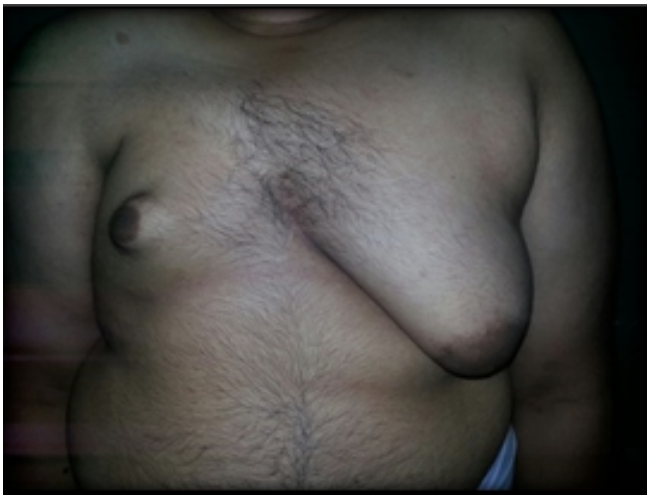

Figure 1. Before Surgery – marked chest wall asymmetry

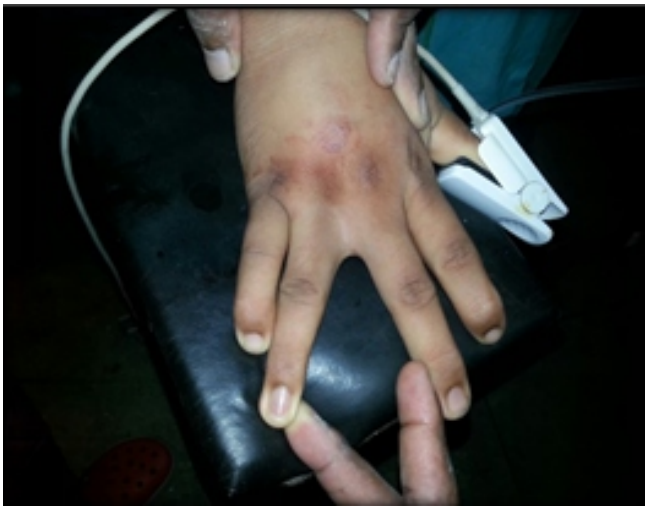

Figure 2. R/S partial syndactyly

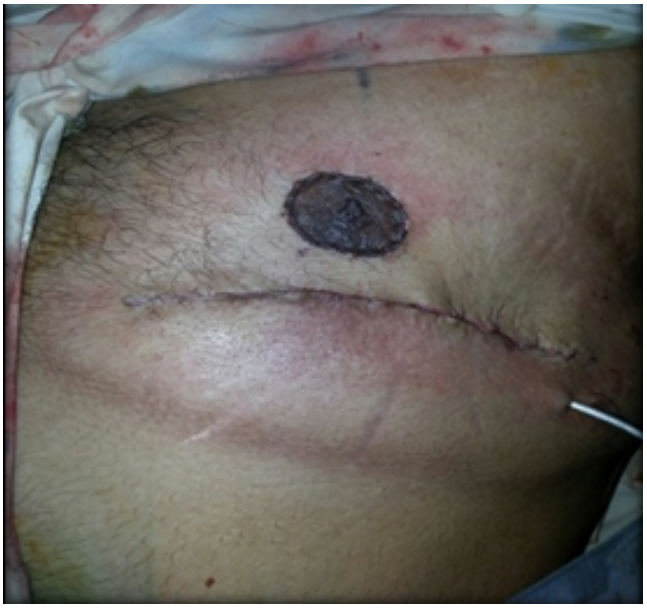

Figure 3. After Surgery

The chest wall in a severely affected individual will have a hairless chest with thinned out subcutaneous tissues with smaller or rudimentary nipple and areolar complex. Underlying ribs may be smaller and deformed. The combined length of the affected upper limb is shorter due to arm, forearm wrist hand shortening. Other finger anomalies are syndactyly or webbed fingers and shorter fingers. The small bone union may lead to what is known as symphalangism. More severe cases may have deficient thoracic muscles (serratus, external oblique, pectoralis minor, latissimus dorsi, infraspinatus and supraspinatus muscles). The lung may also herniate due to hypoplastic or absent anterior ribs (4).

Poland syndrome affected cases are sporadic in occurrence. However, some rare cases show autosomal dominant inheritance pattern. The responsible genes are yet to be determined (5). Management depends on the severity of deformities. In minor chest wall asymmetry, permanent tissue expanders can be used. In severe cases with no underlying rib anomalies can have lattisimus dorsi flap reconstruction. However, if severe, associated rib abnormalities may need to be treated to optimize the eventual outcome (4).

This case was managed surgically to correct the gynecomastia first and lipofilling of the Poland syndrome side subsequently. Although the implant is indicated to improve the cosmesis his poor glycaemic control and obesity precluded from us to embark on this at this point of time.

The surgical reconstruction was complex since the patient wanted to preserve the nipple-areolar complex. What we is a modified version of the wise pattern technique. The only difference is that the nipple-areolar complex is kept on inferiority based pedicle and brought on to the skin through the skin similar to a buttonhole opening to match the future nipple level. This is placed at a similar point in the fourth intercostal space. This is done as the hypoplastic NAC is expected to be moved with the reconstruction of that side. Pre-op and the post-operative photos are provided.

Conclusion

The occurrence of Poland syndrome with contralateral gynecomastia is extremely rare. Satisfactory results for gynecomastia can be achieved by liposuction and nipple-sparing gynecomastia correction surgery.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Poland syndrome is rare but needs to be excluded in patients with congenital hand problems.
- Gynaecomastia correction in grade 4 disease may need to employ techniques used in reduction mammoplasty.