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Seizure Disorder in a Case of Poland Syndrome an Unusual Association

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Abstract: ***Poland syndrome is rare congenital anomaly.it is characterised by absence unilateral chestwall muscle specifically pectoralis major muscle and symbrachydactyly (abnormally short and webbed finger).We report a 4 and ½ years age boy presented with typical features of Poland syndrome and seizure..However, the association of seizure disorder with Poland sequence has not been observed so far. Such an association is being reported here .***

Keywords: Poland Syndrome, Pectoralis Hypoplasia, Seizure

1. Introduction

Poland syndrome consists of ipsilateral hypoplasia or abscent of sternocostal portion of pectoral major muscles with associated hand defect. [(1)]. Others variable associated features are underdevelopment or absence of one nipple including the areola and/or patchy absence of hair in the axilla. [(1,2).] Though very difficult to estimate but current incidence rate is 1 in 7,000 to 1,00,000 live birth.males are affected more (male female ratio 2:1 to 3:1).About 75% cases involve right hemithorax. familial cases are rare with no sex and side prediliction. [(1,2,3),]

We here report seizure in poland syndrome 1st in india and 2nd with world literature.[(4)]

2. Case

A four (4) and half (1/2) years boy presented to opd with complain of generalized tonic clonic convulsion for last 6 months. He was put on oral valparin outside for last 2 months and with no further attack. He was born out of nonconsangunious marriage at term by normal vaginal delivery at home, antenatal, intranatal and postnatal period were uneventful. He achieved all the developmental milestones in times. He had two (2) siblings. None of the family member had similar problem. On examination height 104 cm (>15th percentile), weight 16 kg (>15th percentile) and head circumference 50 cm (< 15th percentile).

His vitals were within normal limit. All peripheral pulses were palpable, right sided chest was flat compared to left side, absent of areola, nipple along with abscent sternal head of right sided pectoralis muscle,(Figure 1) .There were right sided brachydactyly, (figure 2).Systemic examination, eye examination revealed no abnormality.

His serum biochemistry revealed Na-134 meq/l,K-3.5 meq/l,Ca 1.1 meq/l.EEG showed- stage iii and iv NREM sleep record.MRI brain couldnot be done due to financial constrain. He is now asymptomatic on follow up.

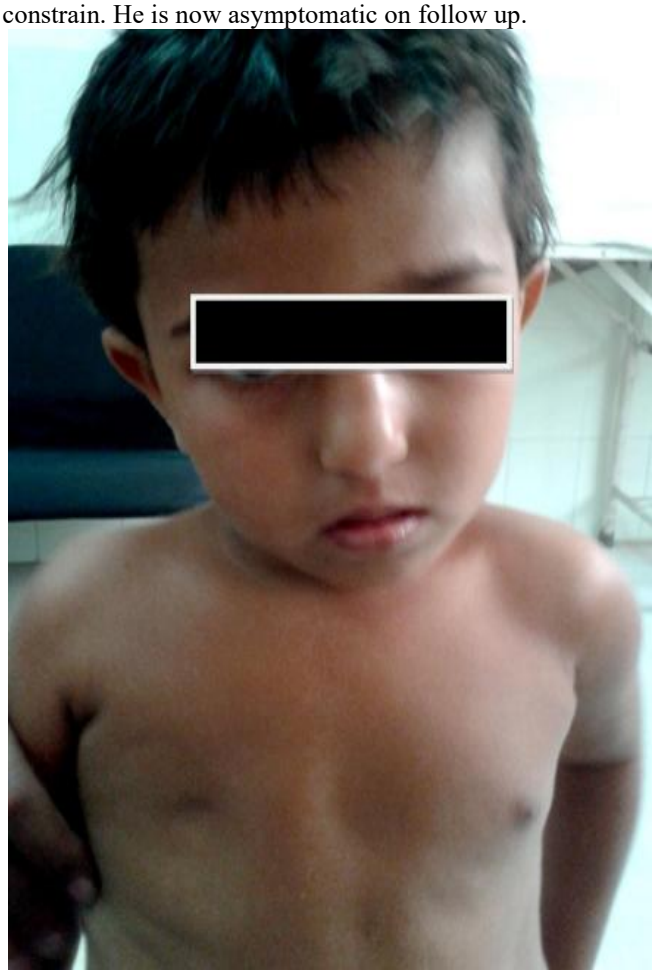

Figure 1: Flat Right Chest, Abscent Nipple and Areola,Abscent Pectoralis Major

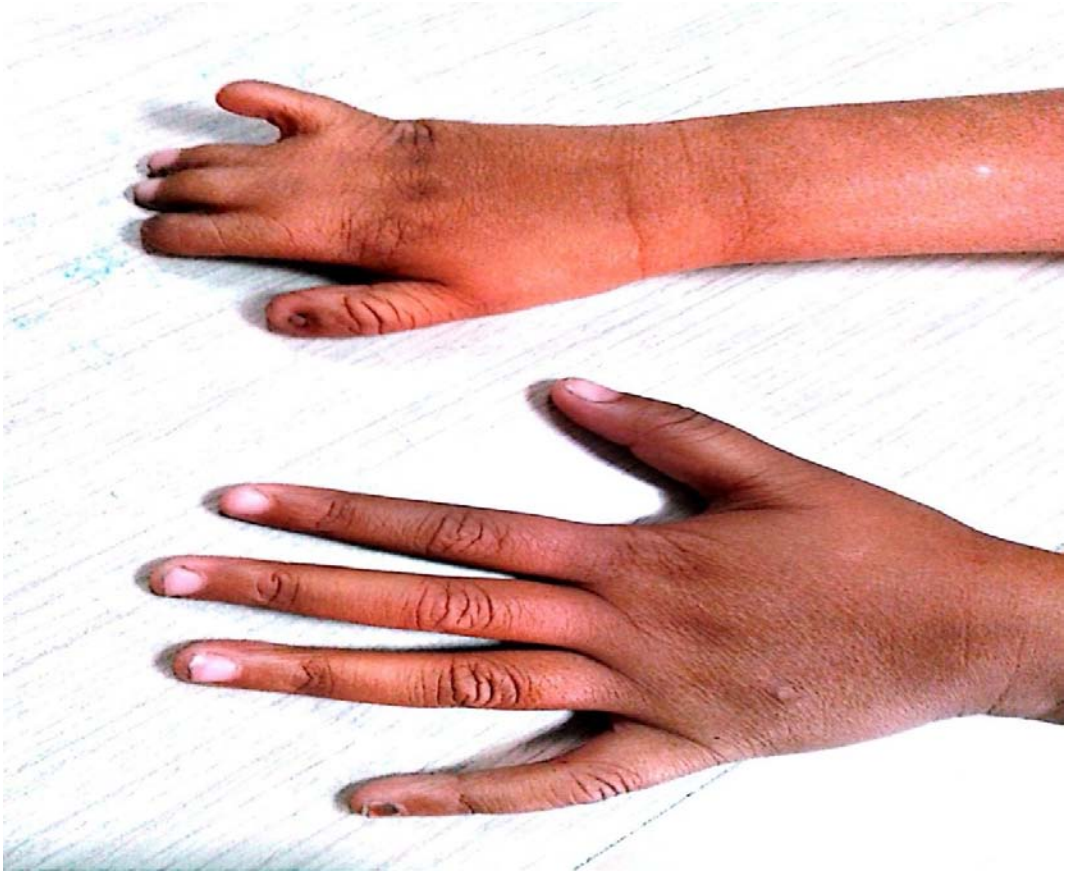

Figure 2: Right Sided Symbrachydactyly

3. Discussion

Dr Alfred Poland got the credit of describing first case of poland anomaly or sequence in cadaver examination in 1841 at Guy's hospital.[5]In his writtings he specifically mentioned the absence of sternal portion of pectoralis major with intact clavicular origin along with absent pectoralis minor, hypoplastic serratus anterior and external oblique muscle. Neither breast hypopasia or hand deformity were included in original description. The definition has under gone evolution and now thought to include shoulder-girdle anomalies with or without upper extremity involvement[6] .Now compulsory diagnostic criterion of this syndrome is the presence of aplasia or hypoplasia of the pectoralis major muscle and at least one of the following combined abnormality - costal aplasia/hypoplasia, depressions of the chest wall, athelia or amastia, absence of axillary hair, hypoplasia of subcutaneous fat, radius hypoplasia and hand anomalies. The syndrome is sporadic in origin with male preponderance.bilateral and lower limb involvement are also noted.[7]

Exact etiopathogenesis is unknown but hypothesis is that syndrome is a result of interruption of early embryonic blood supply of chest wall by subclavian artery branches by thrombus or emboli.[8] It is also called subclavian artery supply disruption sequence (sasds).but it can't explain all the facets.

Though neural defects has been described previously but isolated seizure not documented.[4,9] A neuro imaging may be confirmatory but absence of other features like delayed development, abnormal head circumference, prolonged course with features of raised intracranial tension, dyselectrolytemia points towards seizure disorder.EEG is also corroborative. It may be an accidental association or some vascular pathology may play arole as poland syndrome has a vascular origin. But prognosis of seizure is good. Future report can shed light on this phenomenon.

The patient responded to oral valparin very well and there is no breakthrough seizure. Regarding chest wall reconstruction may be done using the lattisimus dorsi muscle, custom-made silicone prosthesis and/or a breast prosthesis.[10]

4. Conclusion

Poland syndrome very rarely may present with seizure disoredr. Exact etiology is unknown .But prognosis is very good.

5. Acknowledgement

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We expend from my personal account.

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