POLAND'S SYNDROME; CASE REPORT

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INTRODUCTION

Syndrome named after Sir Alfred Poland is described as an absence or under development of the chest muscle (pectoralis) on one side of the body as well as webbing of the fingers (cutaneous syndactyly) on the hand of the same side. Poland syndrome is usually sporadic, although rare familial cases have been described[1]. The incidence of Poland's syndrome varies between groups (male versus female patients and congenital versus familial cases) and ranges from 1 in 7,000 to 1 in 100,000 live births[2]. The cause of Poland syndrome is unknown. However, an interruption of the embryonic blood supply to the arteries that lie under the collar bone (subclavian arteries) at about the 46th day of embryonic development is the prevailing theory[3].

CASE REPORT

A 14 years old female was referred to radiology department for evaluation of under development of right breast. On CT and MRI images the absence of pectoral muscle was noted on rightside. Right breast tissue was also rudimentary along with hypo plastic 2nd, 3rd and 4th ribs. Their costochondral junctions were absent. The sternum was short in length, and showing pectus carinatum. However, no dextrocardia was seen. Right scapula was medially placed and was slightly smaller in size. There was no evidence of syndactyly (webbing). The patient was diagnosed as Poland's syndrome. Patient and her parents were explained about the diagnosed syndrome and referred to plastic surgeon for muscle flap transposition combined with breast augmentation.

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DISCUSSION

Poland's syndrome is a rare congenital anomaly characterized by hypoplasia of the breast and nipple, scarcity of subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, lack of the pectoralis minor and major muscle, aplasia or deformity of the costal cartilages or ribs II to IV or III to V, alopecia of the axillary and mammary region, and unilateral brachysyndactyly. Frequent signs are hypoplastic/absent nipples, scapular anomaly, absent pectoral muscles, brachydactyly (short fingers), dextrocardia, diaphragmatic hernia/defect, syndactyly of fingers (webbing), ulna absent /abnormal, upper limb asymmetry and abnormal rib.

Whenever, there is a large defect of the ribs and lung herniation, the chest wall should be stabilized by using subperiostially split rib grafts from the unaffected side, other bony allo- or auto grafts, mesh-patch, or a combination of several of the above. The construction of the ribs and cartilages not only allows the cosmetic correction of the defect, but also prevents paradoxical respiration and provides firm support for a muscle flap or for a prosthetic implant[4]. Hypoplastic breasts may be augmented using prosthetic implants or musculo-cutaneous flaps . This should be done only after puberty to adapt it to the size of the contralateral, fully developed breast. There are also reports of using upper gluteal flaps or micro vascular-free transverse rectus abdominis muscle (TRAM) flaps nourished by the internal thoracic vessels as recipient vessels, in lieu of the hypo plastic latissimus dorsi[5,6]. Preoperative angiography is helpful to provide assessment of the vascular supply before micro vascular tissue transfer[7].

Separation of the fingers should be performed early, preferably in the first year of life, before abnormal function patterns have

developed and the deformity has progressed. Restoration of some hand function in patients with ectrodactylia is possible by toe-transfer or by prosthesis. Parents must be informed that they should expect the hand to remain hypoplastic[8].

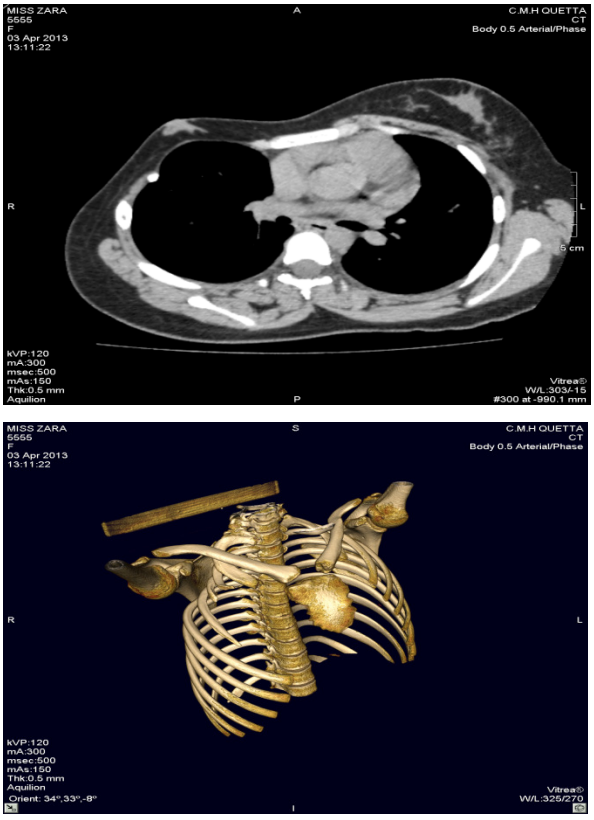

Figure-1: Axial and coronal CT slices of chest with 3D reformatting showing absence of right pectoralis muscles, aplasia of breast tissue on right and hypoplastic upper ribs. Skeletal deformities of scapula and sternum are also seen.

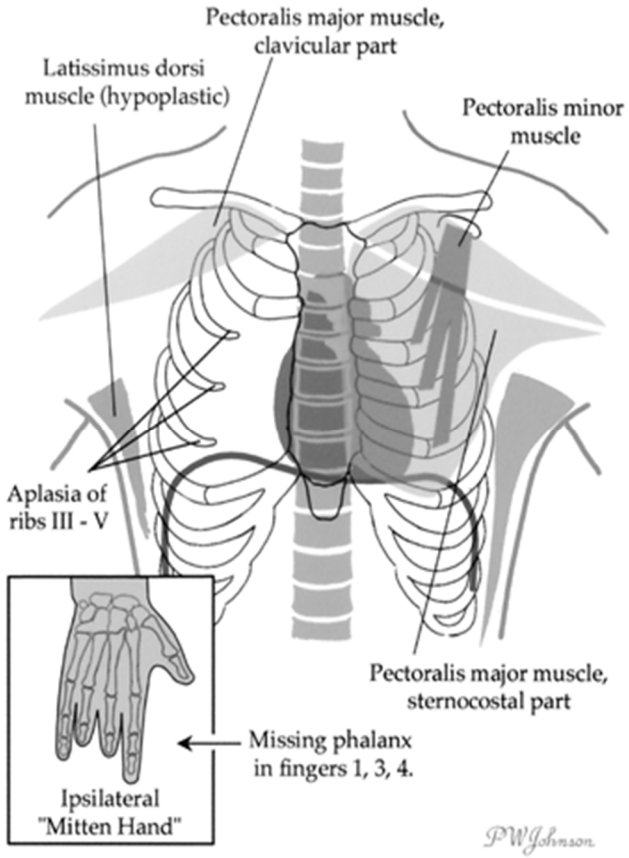

Figure-2: Schematic diagram showing basic features of the Poland's syndrome

REFERENCES

1. Samuels TH, Haider MA, Kirkbride P. Poland's syndrome: a mammographic presentation. AJR Am J Roentgenol 1996; 166 (2): 347-8.
2. Alexander A, Fokin, Robicsek F. (The Society of Thoracic Surgeons) Poland's syndrome revisited. Ann Thorac Surg 2002; 74: 2218-25.
3. Poullin P, Toussirot E, Schiano A, Serratrice G. "[Complete and dissociated forms of Poland's syndrome (5 cases)]". Rev Rhum Mal Osteoartic 1992; 59 (2): 114–20.
4. Glicenstein J. Correction des anomalies thoraciques du syndrome de Poland. Revue généraleet à propos de 20 patients. Ann Chir Plast Esthét 2001; 46: 640-51.
5. Fujino T, Harashima T, Aaoyagi F. Reconstruction for aplasia of the breast and pectoralis region by microvascular transfer of a free flap from the buttock. Plast Reconstr Surg 1975; 56: 178.
6. Tvrdek M, Kletensky J, Svoboda S. Aplasia of the breast—reconstruction using a free tram flap. Acta Chir Plast 2001; 43: 39-41.
7. Beer GM, Kompatscher P, Hergan K. Poland's syndrome and vascular malformations. Br J Plast Surg 1996; 49: 482-84.
8. Ireland DC, Takayama N, Flatt AE. Poland's syndrome. J Bone Joint Surg 1976; 58: 52-8.