CLINICAL QUIZ (p46-47) ANSWER

Figure 1 shows chest asymmetry with hypoplastic left pectoralis muscles and left nipple while Figure 2 shows brachysyndactyly of his left hand. In Figures 3 & 4, there is dextrocardia, absent pectoral musculature on the left side, without abnormalities of ribs and sternum. Other examination of the child, including neurological examination, was normal, and this excludes Moebius syndrome and Klippel-Feil syndrome that are commonly associated with Poland syndrome.

Poland syndrome, a rare congenital syndrome, is usually characterised by the absence or hypoplasia of the breast or nipple, unilateral absence of pectoralis major muscle and varying degrees of congenital upper extremity deformities.[1] The syndrome is usually sporadic but families with an autosomal dominant mode of inheritance have been reported.[2] The incidence is estimated to be 1/30000 and 1/32000.[3] It is more likely to have been on the right side of the body, and is more common in males than females.[1,3] The underlying etiology is unknown but is hypothesised to be related to the interruption of subclavian blood flow in early embryonic period.[4] The clinical synopsis of Poland syndrome is summarised in Table 1.[5-7] Commonly the muscle agenesis in Poland syndrome involves pectoralis major and minor muscles,[8] which is easily diagnosed by physical examination. It may be accompanied by the absence/hypoplasia of the ipsilateral nipple and the absence of the cartilage sections of 2nd/3rd /4th or 3rd/4th/5th ribs.[8,9] Computed tomography and chest X-ray are useful to detect any associated chest wall deformities. In 2001, Al-Qattan et al classified the hand anomalies associated with Poland syndrome[7] and our patient's hand anomaly belongs to type 3 according to this classification. Individual patients have been reported to have tumour/malignancy, but the risk of occurrence is still unknown.

Interestingly most patients with Poland syndrome have right sided involvement but our patient is affected at his left side. Fraser et al reported that 23% of patients with left side affected had dextrocardia[10] whereas in general dextrocardia is found in only ~11% of patients with Poland syndrome regardless of the side of involvement.[9]

The treatment of Poland syndrome is limited to cosmetic operations of chest wall defects especially the pectoral area, and the hand anomalies.

Table 1 Malformations that may co-exist with Poland syndrome

System	Malformations
Thoracic	Pectoral muscle hypoplasia, other thorax muscles hypoplasia, breast abnormalities (30%), thoracic depression, and/or paradoxical motion in the thorax (11-25%), abnormalities of the ribs, clavicle and sternum, Sprengel's deformity, pectus excavatum and carinatum, thoracic teratoma, pleural fibroma, lung herniation (8%)
Skeletal system	Upper limb abnormalities (hypoplasia and brachysyndactyly) (13-56%), gluteal hypoplasia, popliteal webs, club foot, toe syndactyly
Cardiovascular	Dextrocardia, atrial septal defect
Craniofacial	Mandibular prognathism, craniofrontonasal dysplasia, epicanthus, strabismus, ptosis, external ear anomalies, seizures
Gastrointestinal	Hernia, ulcerative colitis, pyloric stenosis, liver herniation
Genitourinary	Renal agenesis, ureteral reflux, undescended testes
Spinal	Scoliosis, hemivertebrae
Skin/hair	Absent axillary hair, skin dimples, hairy naevus, Adams-Oliver syndrome (curls aplasia)
Blood/lymphatic	Thrombocytopenia, leukaemia, lymphoma, spherocytosis

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