

CASE REPORT

POLAND SYNDROME IN A NIGER DELTA WOMAN. REPORT OF A CASE.

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INTRODUCTION

Poland's syndrome was first described following a cadaver dissection by Sir Alfred Poland in 1841, as a chest wall deformity showing absence of the sternocostal origin of the pectoralis major, pectoralis minor as well as hypoplastic serratus anterior and external oblique muscles.¹ However other defining features like breast hypoplasia/aplasia, nipple asymmetry, absence of axillary hair and hand deformities which were subsequently observed, were not noted.² Severe presentations of thoracic skeletal deformities like absence of some anterior and costal ribs, lung herniation, winging of the scapula have also been reported. The condition has been ascribed to the prevailing theory of a vascular development anomaly occurring in the sixth gestation week leading to developmental anomaly of the subclavian artery or its branches.² Incidence ranging between 1 in 7,000 and 1 in 100,000 has been reported among demographic groups (gender, familial or sporadic).² Workers have defined other characterising conditions to include absence of subcutaneous tissue, brachysyndactyly, unilateral absence of axillary and mammary hair.³ Nevertheless, extreme variability has

been noted in its presentation.⁴

We report a case of Poland's syndrome seen in a 21 year old lady in Delta, Nigeria.

CASE PRESENTATION

A 21 year old lady presented with an absence of the left breast. She had lived with this condition for as long as she could remember. General examination was unremarkable. There was no observable thoracic or ipsilateral upper limb deformity. The finger digits appeared normal. Breast examination showed a hypoplastic right breast and nipple, but a fully developed left breast. The right nipple/areola was hypoplastic and elevated in comparison to the contralateral side. There was no family history of a similar condition. Subclavicular or anterior axillary fold absence was not demonstrated. Musculoskeletal examination did not show any bony deformity in the limbs or rib cage. Chest xray showed a well formed rib cage with costal components. However an absent right breast shadow was noted.

DISCUSSION

Poland syndrome is an uncommon occurrence. Although our case occurred in a young female patient, a female /male ratio at 1:2 -1:3 has been described.⁵ Often times, the female patient as in our case seeks cosmetic surgery for her breast asymmetry being a constant feature in female 'Poland patients'. This milder form of Poland syndrome which our case presented with has been described as a

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Clinical photograph showing right breast/nipple hypoplasia
Chest xray showing intact rib cage with absent right breast shadow.

partial Poland sequence with a reported incidence of 1 in 16,500 live births. Our case had no family history of the condition, concurring with the largely sporadic nature of its occurrence. However, a positive family history has been reported in at least 20 cases in world literature.⁶ An occurrence has been described in the medical literature of a family that had Poland syndrome in three successive generations.⁷ This unilateral condition has been reported to be right sided in 60-75% of cases as in our case.² An isolated case occurring in a 6 year old with bilateral absence of the pectoralis major, hypoplasia of the breasts and nipples, symmetric chest wall deformity and bilateral hand anomaly has been described in the literature.⁸ However, the term thoracic dysplasia, has been proposed to describe this bilateral case, as Poland syndrome is described as being a unilateral occurrence.^{9,10} Our case presented in adult life being concerned about the cosmetic aspect of a right sided hypoplasia in comparison to the contralateral fully developed breast. A spectrum extending from mild hypoplasia to amastia has been reported in more than one-third of female patients with Poland syndrome.¹¹ Although the right side showed characteristic elevated, hypoplastic nipple and areola, supernumerary nipples were not

present as has been demonstrated in some cases.²

Our case demonstrated a normal rib cage on chest xray, however the breast shadow was unsurprisingly absent. There have been reports of chest deformity being common place in this condition, contributing to cosmetic disfigurement.² This has been determined to be due to hypoplasia of the involved ribs and costal cartilages.² Aplasia involving up to three ribs with severe chest wall depression has been reported to occur in up to a quarter of the cases.¹¹ In the same vein, severe presentations of lung herniation in association with aplasia of anterior ribs and costal cartilages has been described. Reports of an absence of the sternocostal aspect of the pectoralis major muscle, suggests this is a defining feature of this congenital anomaly.² Few reports of invasive ductal carcinoma occurring in the hypoplastic breast suggests that it is susceptible to disease processes of normal breast tissue;¹² hence monitoring for early detection of breast cancer is on course. Other oncologic conditions including leukaemias, lymphomas, cervical cancers, leiomyosarcoma and lung cancers have also been reported in association with Polands syndrome.^{13,14)}

An absence of limb or finger deformities in our case suggests a less severe form of Poland's syndrome; the syndrome being described as a hand and ipsilateral thorax syndrome with varying degrees of brachysyndactyly.¹⁵ Thus the condition has been reported to be extremely variable in severity and manifestation.⁴

Our case presenting in adult life with marked asymmetry from a fully grown contralateral breast would benefit from temporary tissue expander on the affected side and later placement of a permanent saline or silicone implant. There is evidence that strategic placement of tissue expander may achieve lowering of the high nipple areola complex position in addition to enlarging the nipple areola complex.¹⁶ Contralateral breast surgery towards achieving symmetry may be considered. Incremental breast expansion using saline with silicone implant with an attached port would be beneficial for a growing adolescent.

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