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Poland Syndrome: Description of an Atypical Variant

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Abstract. Poland syndrome comprises a unilateral absence of the large pectoral muscle, ipsilateral symbrachydactyly, and occasionally other malformations of the anterior chest wall and breast. The condition is more frequent among males, and usually occurs on the right hemithorax in the unilateral form. The syndrome is believed to be caused by a genetic disorder that reduces the embryonal circulation in the interior chest artery: the stronger the interaction, the more severe the pathology. This article analyzes an unusual pathologic case in which the 17-year-old patient lacked the large pectoral muscle on the left side, but showed no arterial alteration. This case raises questions as to the true pathogenesis of this syndrome.

Key words: Pathogenesis—Plastic surgery—Poland syndrome

Poland syndrome was first described by Alfred Poland in 1841 as a condition characterized by absence, complete or partial, of the pectoralis major muscle; by hypoplasia of the breast, which can be associated with agenesis of the II, III, IV, V ipsilateral costal cartilage; and by athelia, associated in some cases with ipsilateral symbrachydactyly and alopecia of the axillary and mammary ipsilateral regions [3,4]. The clinical picture described by Poland showed a syndromic manifestation of the same disease previously described in 1826 by Lallemand and in 1839 by Froriep, who described the case of a woman with mere thoracic involvement [3]. Therefore, Poland syndrome may occur with different levels of gravity. In previous descriptions, the only constant was the lack of the sternocostal bundle in the pectoralis major muscle. Other authors (Thomson in 1895 and Bing in 1902) have documented the same lack of the pectoralis muscle and a few other deformities (membranous syndactylies of the ipsilateral hand). In 1962, Clarkson coined the term "Poland syndactily" [3].

Currently, it is assumed that Poland syndrome is characterized by a missing sternocostal bundle of the pectoralis muscle. The disorder is sometimes associated with athelia and seldom with additional deformities to the ipsilateral hand. It occurs in complete or incomplete forms (Haller) or with major or minor manifestations (Vanderbussche). Other forms of the syndrome with additional anomalies have been described, characterized by morphologically altered bones and cartilages of the thoracic cage, with sternocostal malformations and herniation of the lung. According to Lauro's proposed, classification, the patient with Poland syndrome lacks the sternocostal bundle in the pectoralis muscle and has mammary hypoplasia, manifests the aforementioned deficiencies as well as costal anomalies, and exhibits complete thoracic malformation with pulmonary herniation.

Poland syndrome is present at an incidence of 1:7,000 to 1:100,000, with a higher frequency among males (ratio, 2:1-3:1). In 75% of the cases, it is located at the right hemithorax in the unilateral form. Poland syndrome is considered a congenital disorder, although less than 1% of patients have family history (shown in 20 patients by deletion of a dominant autosomic gene) [3,5]. It is believed that in embryonic development, during the sixth week of pregnancy, a momentary interruption or reduction in the circulation of the thoracic artery or one of its peripherical ramifications primes the pathogenetic mechanism of the syndrome and results in different degrees of syndrome severity depending on the length and intensity of the vascular interruption [1-3]. Our experience includes cases of "classical" Poland syndrome, in which the affected morphology and functionality of the pectoralis major muscle was diagnosed by instrumental examinations (electromyography, ecography,

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Computed Tomography) [3,6]. Such manifestations, as described in the literature, have always been associated with athelia, and sometimes with defects of the condrocostal articulations.

Our experience also includes a case of particular interest because of the syndrome's unusual manifestations. A male 17-year-old patient came to our attention because of the psychological problems caused by the congenital athelia of the breast symptoms. At the clinical examination, the agenesis of the complex areola-nipple-mammary gland seemed to be the only manifestation of the Poland anomaly. This was a rare expression of the pathology because the affected area, the hemithorax, was not the usual location for the pathology.

Clinical examination and electromyography showed that the pectoralis major muscle was normo-represented and normo-tonic, even in its sternocostal portion, without alterations to the thoracic condrocostal articulations or to the ipsilateral superior limb. There was severe hypoplasia of the anterior dentate muscle and hypoplasia of the pectoralis minor muscle, as seen in the Tc examination of the thorax and in the position of the ipsilateral omplate, which appeared, in standby position, displaced downward and rotated medially with respect to the controlateral side.

The description of this particular clinical case is important not for its therapeutic resolution (reconstruction of the areola-nipple complex through free-skin grafting from the groin region), but for the unusual alterations. Considering the supposed embryogenesis of the affected structures, this unusual case could give rise to new discussions about the etiopathogenesis of the Poland syndrome.

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