Interesting Case Series

Clinical Manifestations of Poland Syndrome

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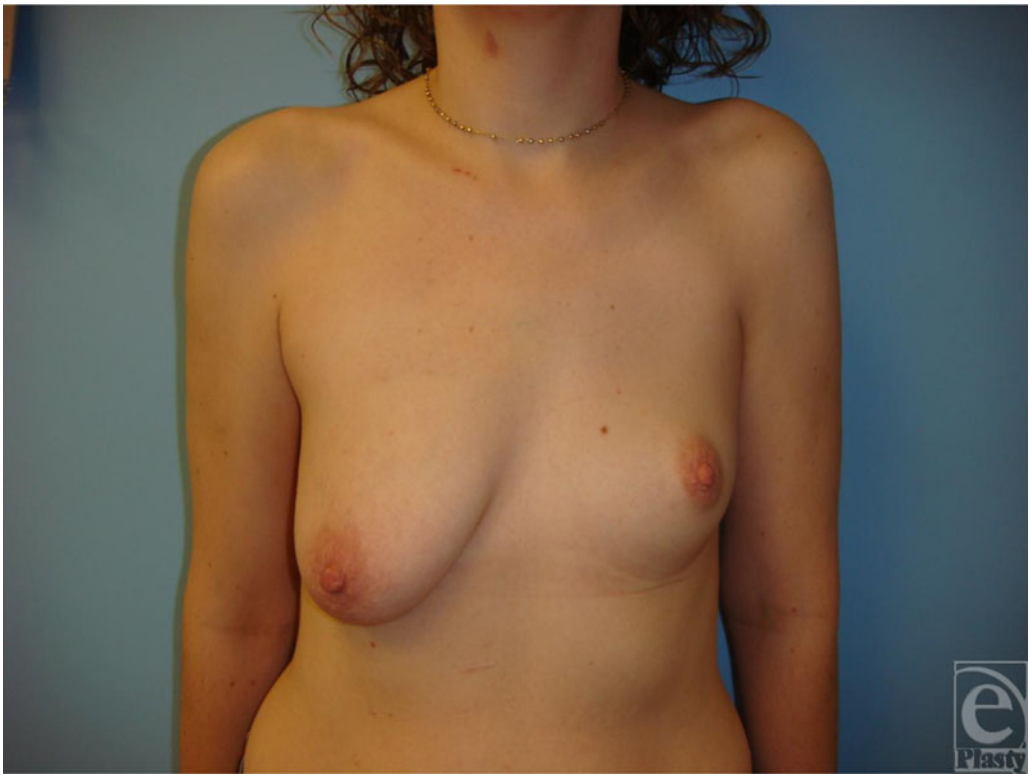

BRIEF DESCRIPTION OF THE PROBLEM

A 28-year-old woman presented with asymmetric breasts, right larger than the left. Previous mammogram showed negative results. She denied mastodynia and had no constitutional symptoms. She complained of not being able to wear her wedding ring on her left ring finger because of short and fused digits.

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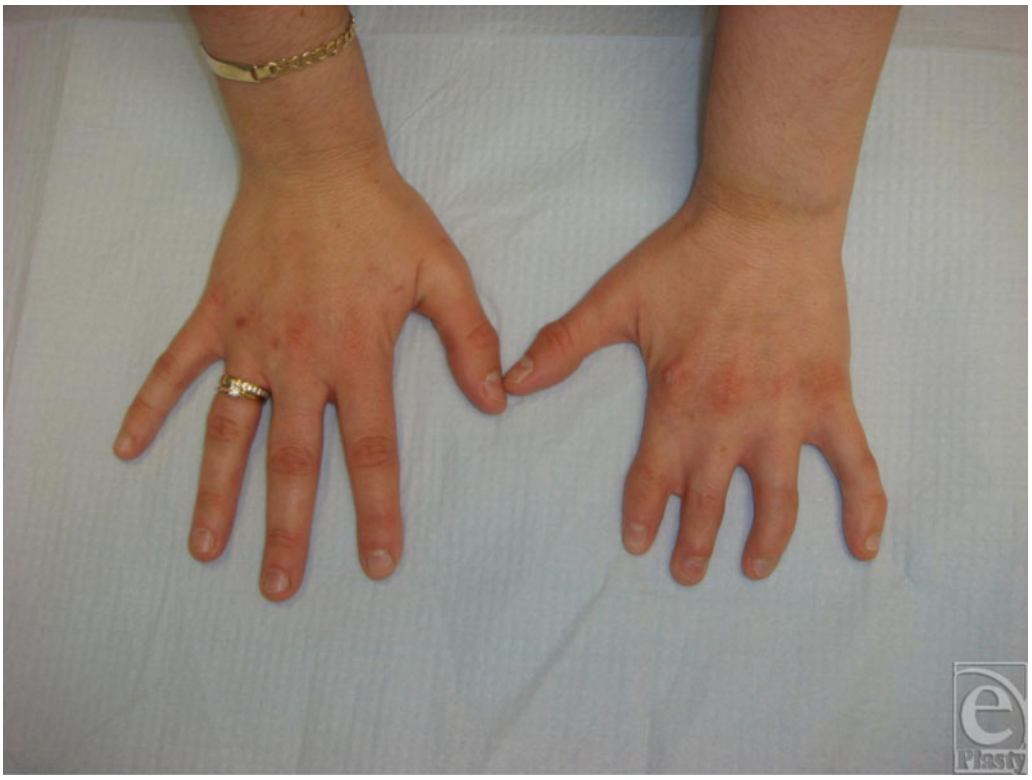

QUESTIONS

1. **What is the incidence and etiology of this condition?**
2. **What structures are involved in this condition?**
3. **What reconstructive considerations must be taken into account pertinent to this patient population?**

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DISCUSSION

This multiparous woman was concerned about the appearance of her left breast and left hand. She kept her hand hidden and wore her wedding ring on her right hand. She presented to the clinic to discuss her condition and reconstructive options.

The differential for this condition includes Poland syndrome and unilateral hypoplasia. Ipsilateral upper extremity involvement suggests Poland syndrome, a rare condition occurring in approximately 1 in 30,000 cases. The right side is more commonly affected, and there appears to be a male predilection. Although the etiology is unclear, it is thought that a vascular accident involving the subclavian artery early in the first trimester may be responsible. Computed tomographic imaging can confirm the diagnosis and delineate involved structures.

Classically, the sternal head of the pectoralis major and pectoralis minor muscles are absent. Supraspinatus, infraspinatus, serratus anterior, and latissimus dorsi may also be hypoplastic or absent. There is a loss of the anterior axillary fold on the affected side. There may also be hypoplasia of the nipple areolar complex, with decreased axillary hair. Hypoplasia or absent ribs are common in more severe cases. Ipsilateral limb involvement includes syndactyly with hypoplastic or absent phalanges, with possible involvement of the radius and the ulna.

Reconstructive options include prosthetic implant placement with ipsilateral latissimus dorsi flap (when present), tissue expansion, external prosthesis, and free tissue transfer. The rectus abdominis muscle is uninvolved in Poland syndrome, and transverse rectus abdominis myocutaneous or deep inferior epigastric artery perforator flaps may be considered. A risk unique to this population of latissimus transposition is postoperative weakening of the shoulder girdle, given the absence of supporting structures. Generally, we wait until after puberty to allow full development of the unaffected breast. While some feel 18 years is appropriate, others initiate tissue expansion at the onset of puberty to facilitate psychosexual development. Because the woman described is ptotic on the right side, she may benefit from mastopexy.

Reconstructive options for simple syndactyly are extensive. Z-plasty is often performed with volar-dorsal interdigitating mirror flaps. Incisions involving the web space should be avoided, and the repair may require full-thickness skin graft interposition. The common digital arteries normally bifurcate proximal to the web space; however, blood supply between the affected digits may be nonduplicated. Therefore, tourniquet should be released at least once during repair to ensure preservation of blood flow after digit separation. For the same reason, separation should remain distal to the arterial bifurcation and 3 adjacent digits should not be separated in 1 stage.

SUGGESTED READINGS

1. Ram AN, Chung KC. Poland's syndrome: current thoughts in the setting of a controversy. *Plast Reconstr Surg*. 2009;123(3):949-53; discussion 954-5.
2. Colombani PM. Preoperative assessment of chest wall deformities. *Semin Thorac Cardiovasc Surg*. 2009;21(1):58-63.
3. Fokin AA, Steuerwald NM, Ahrens WA, Allen KE. Anatomical, histologic, and genetic characteristics of congenital chest wall deformities. *Semin Thorac Cardiovasc Surg*. 2009; 21(1):44-57.

Jonathan Zelken. Clinical Manifestations of Poland Syndrome. www.ePlasty.com, Interesting Case, May 19, 2010

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