Perianal extramammary Paget’s disease affects individuals between the ages of 50 and 80 years and is more common in women and white skin races. Extramammary perianal Paget’s disease (EMPD) is a rare, slow-growing intraepithelial adenocarcinoma that mainly involves the vulvar, perineal, perianal, scrotal, and penile skin.1–3 Skin areas rich in apocrine glands, such as the genital region, are typical sites of EMPD. Signs and symptoms are skin lesions, often mistaken as eczema, that may be itchy or painful. The treatment of Paget’s disease is essentially surgical. After the wide local excision of the lesion with a safe margin, the next step is the reconstruction of the defect. This is often challenging. It is important to restore the shape, function, and volume. Skin grafting is vulnerable to trauma and infection. Given the size of the defect, some local flaps have insufficient volume or are difficult to fit into place. Pedicled perforator flaps are a better option for perineal reconstruction than myocutaneous flaps, as they avoid the use of a large designated vessel and undesirable functional and cosmetic results of the donor site area.4,5 Defects of posterior and lower regions of perineum can be successfully reconstructed by pedicled perforator flaps, based on internal pudendal artery.6 Other options include perforators originating from the perineal or obturator artery. A polygonal shape of the flap can be used to avoid anal stenosis and multiple other complications.

CASE REPORT

A 47-year-old woman was referred to our hospital, with a 4-year history of anal itchiness and an erythematous eczematoid skin rash in the perianal region. The woman complained that the lesion had gradually increased in size and she suffered from severe itching. A perianal shave biopsy revealed an EMPD. The preoperative testing included a complete blood count, colonoscopy, abdominal and chest computed tomography, digital mammography, an abdominal and pelvic ultrasound, and pelvic examination. The test results were negative for cancer. The procedure included a wide local excision, resulting in a semicircular defect of 3.5 cm in diameter. The reconstructive technique involved a pedicle flap based on an internal pudendal skin perforator artery. The perforator was identified (Fig. 3), and the subcutaneous flaps, as they avoid the use of a large designated vessel and undesirable functional and cosmetic results of the donor site area.4,5 Defects of posterior and lower regions of perineum can be successfully reconstructed by pedicled perforator flaps, based on internal pudendal artery.6 Other options include perforators originating from the perineal or obturator artery. A polygonal shape of the flap can be used to avoid anal stenosis and multiple other complications.

From the *Department of Plastic Surgery, St. Savvas Cancer Hospital of Athens, Athens, Greece; †Department of Plastic Surgery, Evangelismos General Hospital of Athens, Athens, Greece; and ‡Department of Plastic Surgery, 401 General Military Hospital of Athens, Athens, Greece.

Received for publication October 5, 2017; accepted January 12, 2018.

Copyright © 2018 The Authors. Published by Wolters Kluwer Health, Inc. on behalf of The American Society of Plastic Surgeons. This is an open access article distributed under the Creative Commons Attribution License 4.0 (CCBY), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

DOI: 10.1097/GOX.0000000000001697
neous vessels. The flap was advanced to cover the defect and sutured in place. The diamond-shaped flap covered the defect fully, with 1 quarter of its surface used to cover the intra-anal portion of the defect. The donor side was closed directly. Six months after surgery, the patient is disease-free and has returned to her activities (work, swimming, jogging). The functional and aesthetic results are successful (Fig. 4).

**DISCUSSION**

Pedicled perforator flaps based on internal pudendal artery or other adjacent arteries, such as perineal, obturator, or inferior gluteal artery, are a better option for perineal reconstruction than myocutaneous flaps of gracilis, vertical rectus abdominis myocutaneous flap, oblique rectus abdominis myocutaneous flap, deep inferior epigastric perforator flap, avoiding the use of a large designated vessel and undesirable functional and cosmetic results of the donor-site area.

The microsurgical dissection technique of internal pudendal perforator flaps for reconstruction of perineal defects is relatively simple. The polygonal diamond shape of the flap is an easy and highly successful method for better flap insetting in the perianal region. The diamond flap allows for the reconstruction of the skin and of the anal mucosa, thus preventing anal stenosis. Furthermore, it allows the surgeon to avoid the curvilinear perianal incision, which often leads to anal stenosis.
A modified V-Y technique, the flap in a diamond-shaped pattern for perineal reconstruction based on internal pudendal artery or other adjacent arteries, such as perineal, obturator, or inferior gluteal artery, is a safe, efficient method that provides maintenance of maximal blood supply to the flap, minimal tension to the suture lines, and allows primary closure of the donor site. The diamond perforator flap is a simple, modern surgical concept, which minimizes local complications (flap necrosis), is easy to apply to the perianal region, and is not associated with anal stenosis. This flap is a modification of V-Y flap, because the typical V-Y flap would cause anal stenosis and could not be applied beyond the sphincter.

Parthena I. Deskoulidi, MD
St. Savvas Cancer Hospital
St. Alexandras Avenue 171
11522 Ampelokipoi
Athens, Greece
E-mail: deskoulidinena@yahoo.gr

REFERENCES
1. Darier J, Coulillaud P. Sur un cas de maladie de Paget de la région périnéo-anale et scrotale. Ann Dermatol Syphiligr. 1893;4:25–31.
2. Zollo JD, Zeitouni NC. The Roswell Park Cancer Institute experience with extramammary Paget’s disease. Br J Dermatol. 2000;142:59–65.
3. Heymann WR. Extramammary Paget’s disease. Clin Dermatol. 1993;11:83–87.
4. Sinna R, Qassemyar Q, Benhaim T, et al. Perforator flaps: a new option in perineal reconstruction. J Plast Reconstr Aesthet Surg. 2010;63:e766–e774.
5. Saint-Cyr M, Schaverien MV, Rohrich RJ. Perforator flaps: history, controversies, physiology, anatomy, and use in reconstruction. Plast Reconstr Surg. 2009;123:132e–145e.
6. Manchot C, Ristic J, Morain WD. The Cutaneous Arteries of the Human Body. New York, N.Y.: Springer-Verlag; 1983.
7. Wei FC, Mardini S. Free-style free flaps. Plast Reconstr Surg. 2004;114:910–916.
8. Caplin DA, Kodner IJ. Repair of anal stricture and mucosal ectropion by simple flap procedures. Dis Colon Rectum. 1986;29:92–94.
9. Anderson KD, Nesman KD, Bond SJ, et al. Diamond flap anoplasty in infants and children with an intractable anal stricture. J Pediatr Surg. 1994;29:1253–1257.
10. Becker DW Jr, Massey FM, McCraw JB. Musculocutaneous flaps in reconstructive pelvic surgery. Obstet Gynecol. 1979;54:178–183.
11. Shukla HS, Hughes LE. The rectus abdominis flap for perineal wounds. Ann R Coll Surg Engl. 1984;66:337–339.
12. Robbins TH. Rectus abdominis myocutaneous flap for breast reconstruction. Aust N Z J Surg. 1979;49:527–530.