2014

Delayed Repair of Myelomeningoceles

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Elsevier Editorial System(tm) for World Neurosurgery
Manuscript Draft

Manuscript Number:

Title: Delayed repair of myelomeningoceles

Article Type: Technical Note

Keywords: myelomeningocele; pediatric neurosurgery; spina bifida; spinal dysraphism

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Abstract: Objective: Myelomeningocele is a defect that is typically surgically repaired within the first few days of life in developed countries to minimize the risk of meningitis. If left unrepaired, these children may survive to have their meningocele sac epithelialize. The surgical reduction and closure of an epithelialized myelomeningocele represents a unique challenge for the neurosurgeon, as it requires a modification of the typical closure technique.
Methods: 10 years experience in 97 patients with the delayed (>6 months) repair of myelomeningoceles was the basis of this report.
Results: Repair technique in a child with a myelomeningocele that was not repaired at birth presented a surgical challenge whose solutions are presented herein.
Conclusion: Delayed closure of myelomeningoceles is facilitated by adherence to lessons learned form surgical experience on medical missions to Guatemala.

Delayed repair of myelomeningoceles

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Abbreviations: VCU = Virginia Commonwealth University; MMC = myelomeningocele; SC = spinal cord; PL = placode

Abstract

Objective: Myelomeningocele is a defect that is typically surgically repaired within the first few days of life in developed countries to minimize the risk of meningitis. If left unrepaired, these children may survive to have their meningocele sac epithelialize. The surgical reduction and closure of an epithelialized myelomeningocele represents a unique challenge for the neurosurgeon, as it requires a modification of the typical closure technique.

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Introduction

Myelomeningoceles affect approximately 2500-6000 children a year in the United States(5). Internationally the rate varies from a low of 0.1 per 1000 in Africans to 12.5 per 1000 among Celts(7). In Guatemala, where we have made our past medical missions, the incidence was 2.34 per 1000 in the year 2000(2). Many children do not have ready access to medical or neurosurgical care. A great many live in remote areas where the mothers have limited resources and may not receive prenatal care or vitamin supplementation. These are the children in whom we see delayed presentations of open neural tube defects during our screening clinic.

Myelomeningoceles are formed by failure of closure of the neural tube dorsally, leaving a malformed open neural placode that fuses with the skin. Most defects are in the thoraco-lumbar region. The etiology is felt to be multifactorial, including a genetic basis, but is strongly linked to hypofunction of a folic acid-dependent process during early embryonic development (week 4). There is an association with other nutritional deficiencies, such as folate and zinc, and some medications, especially anticonvulsants(5). Overall, the incidence worldwide has declined(7). This is attributed in part to improved nutrition, prenatal vitamins, and in some instances to improved prenatal testing.

The neuroectoderm in humans is visible at post-ovulatory day 16. By day 17-19 it has a midline fold called the neural groove that subsequently deepens giving rise to lateral neural folds(5). This process of the formation and closure of the neural tube is called primary neuralation, which takes place over days 18-26. These folds meet and

begin to close on days 21-23, with the anterior neuropore closing first and the posterior neuropore closing after this. It is failure of closure of the posterior neuropore that leads to the development of a myelomeningocele. The normal structures (spinal cord, arachnoid, dura mater, and skin) do develop but simply fail to approximate. .Furthermore, lack of skin and dura closure leaves the neural tissue exposed to the environment.

The primary treatment of myelomeningoceles is early surgical closure of the defect to prevent meningitis and for protection of the neural placode. In addition, closure of the neural placode is performed in hopes of preventing tethering of the exposed surface and an attempt to preserve more proximal spinal cord function by restoring its normal environment. Delayed closure is associated with an increased incidence of meningitis after 72 hours. When access to surgical treatment is unavailable, these children may survive, allowing their myelomeningocele sacs, including the placode, to epithelialize. Lorber reports a 100% mortality if untreated, however some children survived greater than 6 months(4). These patients make up the majority of the children who presented for neurosurgical evaluation and treatment at our clinic in Guatemala. This clinic was sponsored by the Pediatric Foundation of Guatemala and the International Hospital for Children. The delayed presentation of these children required that we adopt techniques that allowed us to reproduce the type of closure that has proven most successful in neonates with MMC's. These techniques are illustrated in this paper. Delayed closure of a small series of epithelialized MMC's has been published previously(3), but it did not specifically address the technical challenges we faced or the solutions that we found most useful.

Patients and Methods

We began seeing patients in Guatemala City in 1996 on an annual or bi-annual basis. Of those patients, we treated 108 children for myelomeningocele. Data was available for 97 of these patients. The average age at treatment was 9.9 months . The age range was from 1 day to 11 years of age. Children under 3 weeks of age (n=4) were excluded form the analysis. There were 13 thoracic and 5 cervical myelomeningocele repairs performed. The remainder were thoraco-lumbar or lumbar sacral myelomeningoceles. We were unable to close 2 of the myelomeningoceles due to the size of the defect. The majority of these children also required concomitant shunting.

Surgical technique

Myelomeningoceles that have not been closed at birth will epithelialize if the child survives. A typical example is represented in Figure 1. Just as with primary closure cases, careful consideration must be given to the size of the lesion and associated bony deformity to judge the need for the ability to perform a primary closure. There is no magic number for the size of the defect that can be closed, as the ability to perform a successful primary closure is multifactorial and includes the infant size, amount of viable skin, and ability to correct associated bone deformity, especially kyphosis. Data from plastic surgery suggest that the minimal sized defect referred for plastic surgery collaboration is 18 cm^2(6).

A standard sterile preparation is performed as epithelialized surface presents no special concerns for iodine intoxication (Figure 1). For children with open MMC, iodine

preparations are avoided for the theoretical risk of iodine-induced hypothyroidism(1) as well as the potential damage to neural tissue. The placode can be identified near the center of the sac, although compared to its perinatal appearance, the epithelialized placode margins are less well-defined defined, looking more like a keloid than exposed neural tissue. An incision is outlined with a sterile marking pen starting in the normal skin midline 1 cm above the sac and extending at the perceived junction of normal skin and epithelialized arachnoid on each side of the sac. Infiltration with ¼ % lidocaine and epinephrine 1:400k solution was used for pain control and hemostasis under the area of normal skin above the sac. Incisions were made into the sac circumferentially, usually at the top of the sack near the junction of the presumed placode and the epithelialized arachnoid, with very careful attention to hemostasis, as significant skin vessels may be encountered. Once the sac was opened, the anatomy became clearer as the proximal spinal cord was then identified rostrally leading to the placode (Figure 2). In all cases, we attempted to preserve nerve roots arising from the placode, though sacrifice of roots, particularly distal ones from the placode, may be done without new deficit.. At this point the surgery typically became quite different from a perinatal repair; the skin was removed from the residual placode by sharp dissection (Figure 3). The technique requires that a plane between skin and placode be found. It was started by gently retracting the top of the ellipse of skin and identifying the neural placode, which will appear gray and friable, whereas the skin that is firm and pale. We developed this plane sharply with a scalpel or sharp scissors until the entire skin ellipse has been freed from the placode. The placode was then be closed (neuralated) suture (we used 5 or 6.0 proline). Inclusion of epithelium into the deep closure risks epidermoid formation in the future. Just as in perinatal repair,

closure of the raw, exposed surface of the placode will help minimize scar formation and diminish the potential for re-tethering.

At this point, closure was more standard. The dural edges were freed from their fusion to the skin by finding the epidural plane rostrally and proceeding circumferentially with a sharp scissors by placing one blade in the epidural space and the other in the sac. The epidural fat was our guide for staying in the correct plane, as the dorsal paraspinal fascia may be a false layer of dissection. The dura was then mobilized toward the spinal cord. Multiple perforating arteries needed to be coagulated and divided to accomplish this, but, in general, no roots needed be sacrificed. The dura was then closed in a water-tight fashion. The skin edges were mobilized as needed by undermining over the fascia. Occasional facial relaxing incisions were used. In several cases of significant bony kyphosis, removal of the kyphus with small rongeurs was required. Rotational skin flaps or skin grafts for larger defects may be needed, but are the exception, not the rule, and we had only two in the series that could not be closed primarily.

Conclusions

Delayed repair of spinal myelomeningoceles is encountered in environments where access to neurosurgical care is limited or impaired. Such a situation has been encountered in our experience of ten years in Guatemala City. The primary goal of immediate, perinatal treatment of MMC's is to prevent meningitis, protect the neural placode, and to prevent spinal cord tethering at the placode. Children that do not have the option of immediate perinatal repair may survive to present for delayed closure and often have large, disfiguring sacs. The surgical repair of these sacs require techniques that

differ from those used for perinatal MMC closure, due to the fact they are epithelialized and may have undergone changes associated with previous infection. MMC closure is indicated to prevent sac rupture, correct deformity, and to untether the spinal cord. The surgical technique of closing these MMC's has been outlined with special attention to the removal of skin from the placode.

Figure legends

Figure 1: 7 months old with an epithelialized MMC. A) photograph of the myelomeningocele; B) artist rendering to emphasize the subtle distinction between placode and epithelialized arachnoid

Figure 2: Intraoperative photograph demonstrating the proximal spinal cord (SC) leading to the placode (PL)

Figure 3: : Intraoperative photographs: A) placode being prepared for sharp dissection of epithelium off of neural tissue, proximal spinal cord (SC) seen leading to the placode; inset B) epithelial layer removed from placode

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Figure 1

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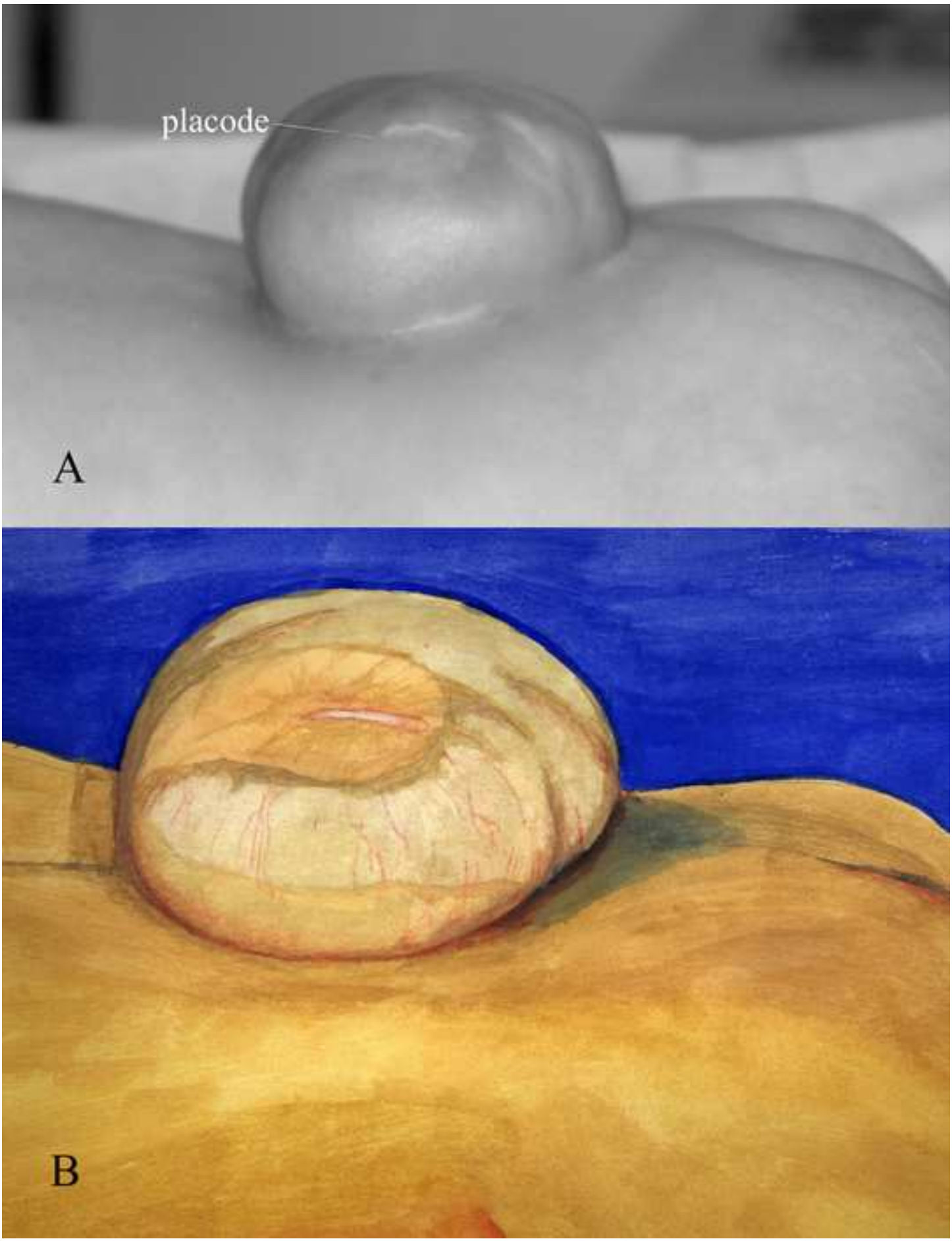

Figure 2

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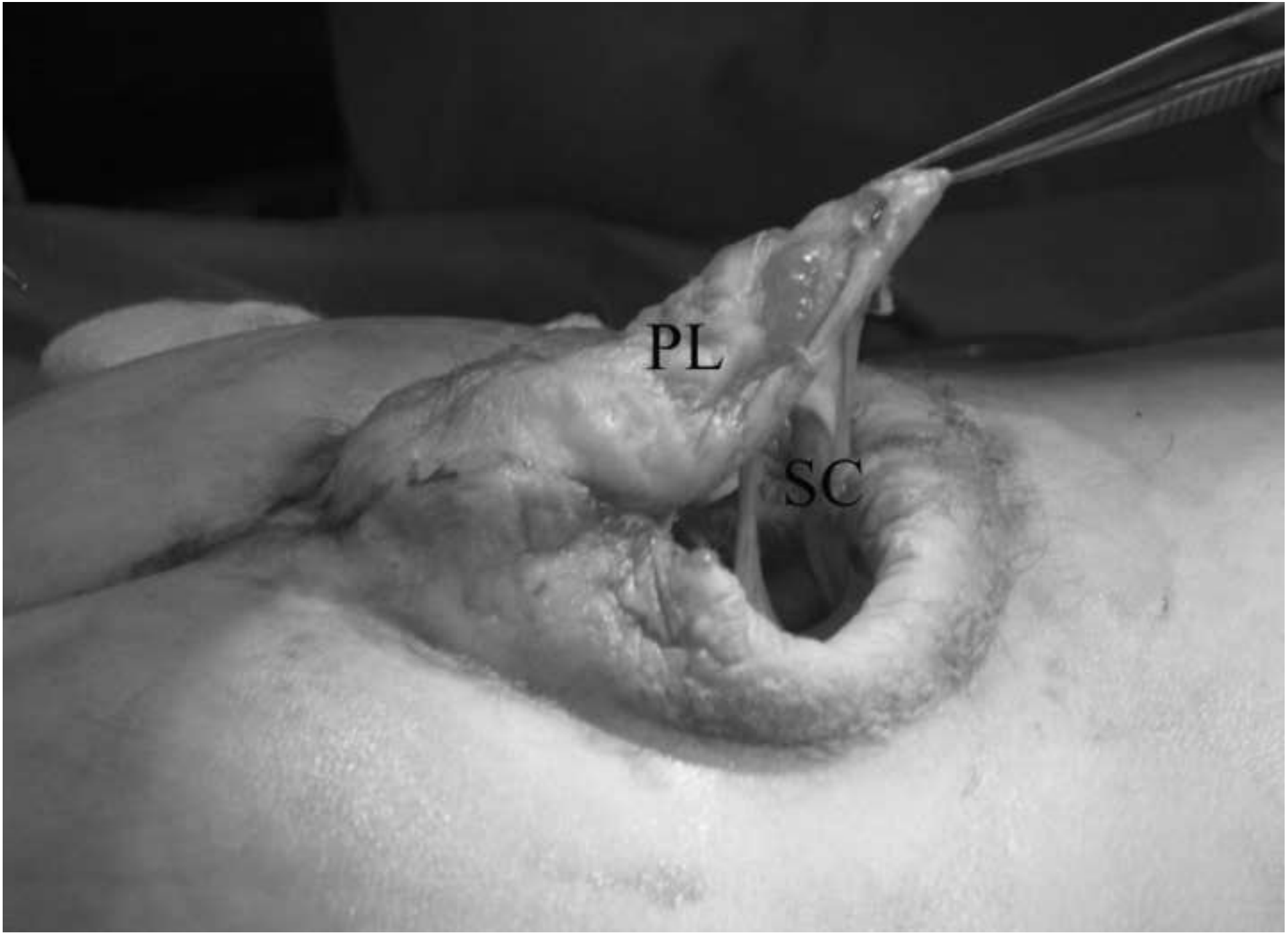

Figure 3

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