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Arthroplasty in patients with rare conditions

Parsonage-Turner Syndrome and Closed-Incision Negative-Pressure Wound Therapy After Total Hip Arthroplasty in a Case of Marfan Syndrome

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ABSTRACT

Negative pressure wound therapy (NPWT) is a postoperative wound care method, which has recently become an ongoing field of research in hip and knee arthroplasty. We report the successful management of wound dehiscence and infection after THA in a case of Marfan syndrome by closed-incision negative-pressure wound therapy (ciNPWT). Our patient also developed a rare postoperative neurologic complication, that is, Parsonage-Turner syndrome (PTS). To our knowledge, this is the first report of PTS and ciNPWT use for SSI after THA in a Marfan patient. As wound dehiscence and infection can occur after THA in Marfan patients, we propose ciNPWT as an option to treat or even prevent (prophylactic use) such complications in this rare group of patients.

Introduction

Negative pressure wound therapy (NPWT) is a postoperative wound care method, which involves placing a spongy foam inside an open or on a closed wound, sealing it by a transparent drape, and connecting it to a suction device [1]. It has shown promising results in fields of plastic, trauma, and orthopedic surgery. The use of NPWT in joint reconstructive surgery has recently become an ongoing research field [2]. Despite the scarcity of high-ranking evidence, significant findings favor NPWT use after hip and knee arthroplasty [3]. The most robust available evidence supports the prophylactic use of NPWT in high-risk patients after arthroplasty [4-7]. However, NPWT use as an adjunct in treating acute periprosthetic joint infection (PJI) is supported only by a few case-series [8-10].

Marfan syndrome (MFS) is an autosomal-dominant connective tissue disorder caused mainly by fibrillin-1 gene mutation, which is the major constituent of microfibrils and elastic fibers, hence determining the elasticity and structural sufficiency of connective tissue [11,12].

Parsonage-Turner syndrome (PTS), or brachial plexus neuritis, is an inflammatory idiopathic brachial plexopathy characterized by multifocal involvement of brachial plexus nerves, motor more than sensory, which occurs after a predisposing condition such as surgery in nearly half the patients [13-15].

The study aims to report the successful use of closed-incision negative-pressure wound therapy (ciNPWT) in managing surgical site infection after total hip arthroplasty in a patient with Marfan syndrome, who was further complicated with PTS. To the best of our knowledge, this is the first report of ciNPWT use and PTS after THA in Marfan patients.

Case history

A 26-year-old female, known case of Marfan syndrome, came first to our hip subspecialty orthopedic clinic with complaints of limping and bilateral hip pain (Fig. 1). She kindly gave her informed consent for her story to be presented and discussed in this report. The patient had a history of bilateral Salter pelvic and femoral shortening osteotomies due to DDH with no other medical comorbidities. On examination, there was a notable limitation of motion in both (left/right) hips with flexion of 80°/100°, abduction

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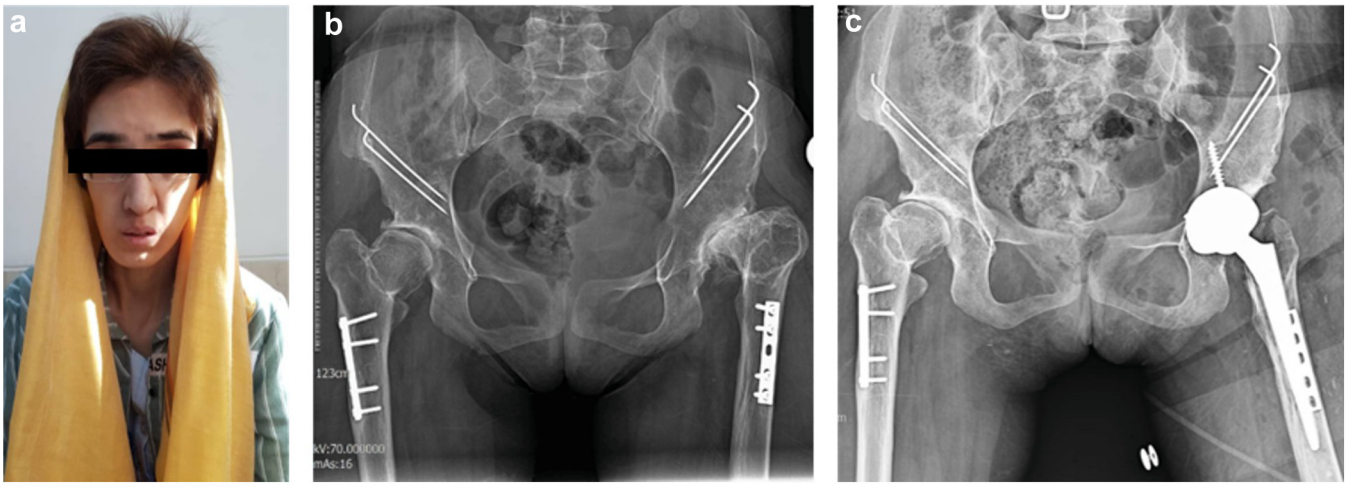

Figure 1. The patient's facies (a) and preopertaive (b) and postoperative pelvic radiographs (c).

of 20°/20°, internal rotation of 15°/35°, and external rotation of 20°/40°, respectively. Considering severe degenerative joint disease (DJD) of both hips, more severe on the left, she became a candidate for left total hip arthroplasty (THA). She had a preoperative Charlson Weighted Comorbidity Index of zero and American Society of Anesthesiologists (ASA) physical status score of one. Her body mass index was 15.6 (height = 166 cm, weight = 43 Kg). The left THA was performed through a direct anterior approach in the supine position with the patient's arms placed on well-padded armboards in neutral abduction. The prosthesis components of use were Wagner Cone stem (16 mm), Continuum shell (46 mm O.D.), VerSys femoral head (28 mm, +0 neck length), and Longevity liner (28 mm I.D.) from Zimmer-Biomet Co. The wound was closed by proper suturing of deep fascia, subcutaneous tissue, and skin and finally covered by a classic occlusive sterile dressing. Not a few hours after surgery, the hip prosthesis was dislocated while taking a postoperative portable radiograph. She underwent open reduction surgery the day after, and a hip-abduction brace was administered thenceforth. She walked successfully using the brace and a walking frame the next day.

On POD[1] four, the serosanguinous discharge was reported from the surgical site. The sterile dressing was then changed regularly; however, secretion was not ceased and even further increased, so that wound dehiscence finally occurred (Fig. 2a). The patient underwent irrigation, debridement, and wound closure surgery on POD 21, and deep wound tissue was sent for bacterial culture. After 3 days, the dressing was soaked with secretions again (Fig. 2b). The culture was found positive for multi-drug-resistant Acinetobacter *baumannii* (resistant to cephalosporins, ciprofloxacin, imipenem, and ampicillin-sulbactam). Based on the infectious disease specialist's consult, the patient was consequently treated by Meropenem and Colistin for 10 days (POD 26-36). However, the wound continued to discharge and, despite proper wound care and antibiotic therapy, dehisced again on POD 30 (Fig. 2c).

Meanwhile, what complicated the patient's management was the unexpected occurrence of biochemical disturbances and neurologic deficits. It started by facial paresthesia, bilateral shoulder pain, and upper limb weakness, more severe on the right, on POD 26, to which was further added hypophonia, lower limb weakness, a patchy sensory deficit of upper limbs, and tetanic spells. She lost her ability to walk due to muscular weakness. The neurologic examination of the patient's extremities is shown in Table 1. The plantar reflexes were bilaterally equivocal, and the deep tendon reflexes of upper and lower limbs were 0-1^{+} on both sides. The patient was subsequently admitted to the ICU. She had intractable hypocalcemia, hypokalemia, and hypomagnesemia, which took more than 10 days to control under the internal medicine specialist's supervision. The facial paresthesia and tetanic spells were controlled after hypocalcemia and hypomagnesemia correction; however, other neurologic deficits persisted, which fostered full neurologic assessment based on neurologic consult. Brain and spinal MRI showed no significant findings. Nevertheless, full four-extremity and para-spinal EMG-NCV indicated bilateral early subacute brachial plexopathy (right upper trunk plexopathy and left lateral/ posterior cord plexopathy) and left subacute lumbosacral plexopathy. Laryngeal stroboscopy revealed left true vocal cord paralysis. The CSF analysis was nonproductive. The immunologic laboratory profile of the patient is shown in Table 2. Bilateral brachial plexus MRI with gadolinium revealed the increased thickness of left C6-8 and right C4-7 nerve roots in favor of an inflammatory process.

According to the expert neurologist's opinion, lumbosacral plexopathy reported in EMG-NCV could be explained by recent THA, but bilateral asymmetric subacute brachial plexopathy together with bilateral shoulder pain, asymmetric motor and patchy sensory involvement of upper limbs, and increased immunologic markers were in favor of an autoimmune process mainly brachial plexus neuritis, or Parsonage-Turner syndrome. She received a course of IVIG under neurologic consult, which improved her sensorimotor deficits. Finally, she was able to walk again using a walking frame. The muscle forces and sensory deficits were improved; however, some degree of deficit remained.

The complicated state of the patient postponed definitive surgical management of the dehisced wound until POD 46. We performed irrigation and debridement and closed the wound completely. However, instead of classic occlusive dressings, negative-pressure wound dressing (NPWD) was applied this time in the operation room right after wound closure. We used the vacuum V.A.C. device (FAPSCO., Isfahan, IRI) for NPWD with a 100-140 mmHg negative pressure, exchanged and set at the discrete of a wound expert every 3 days (Fig. 2d). The result was unbelievable after 3 weeks, with the wound almost healed and no sign of previous discharge and dehiscence (Fig. 2e). Subsequently, we

[1] Postoperative days are reported from primary surgery day.

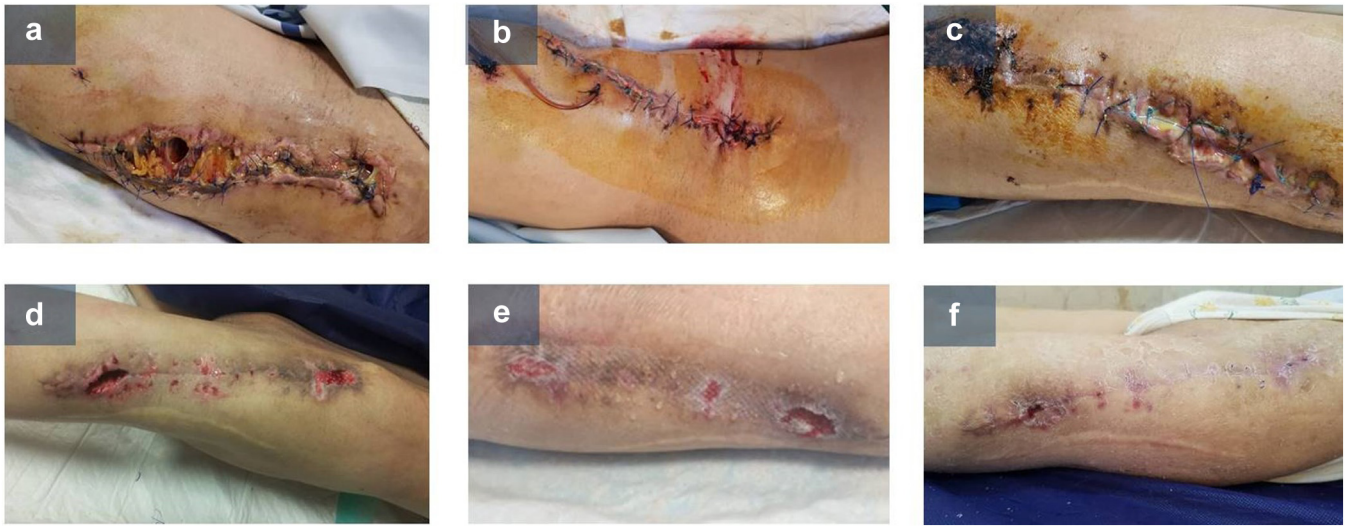

Figure 2. The patient's left hip wound through different stages of treatment: (a) severe dehiscence 4 days before the first session of I&D (POD 17); (b) discharge from proximal end of the wound 3 days after first I&D (POD 24); (c) recurrent wound dehiscence 5 days before the second I&D (POD 41); (d) significant response to negative-pressure wound therapy 8 days after the second I&D and NPWD application (POD 54); (e) complete wound closure at the time of patient's discharge (POD 69); (f) full wound healing 2 weeks after discharge (POD 83).

discontinued the NPWD and applied a sterile occlusive dressing. She was finally discharged after 70 days of admission with the recommendation of regular sterile change of the dressing at home.

At 2-week follow-up, the wound was completely healed (Fig. 2f). Her general condition was normal, and she walked properly with a walking frame. The proximal and distal muscle forces of four limbs were at least 4/5 with some sensory deficits in the volar surface of three radial fingers. She was also under close surveillance in the neurology clinic.

After 26 months, we contacted the patient and asked her for a follow-up visit. She mentioned no problem with the wound to date. On examination, no complication was seen (Fig. 3). She walked independently without an assisting device. However, the muscle forces of upper limb, mostly the thumb pinch and hand grip, were 4/5, and she had problem in activities involving these muscles. Other muscle forces were normal. Some residual sensory deficits also remained in the volar surface of thumb and index fingers. We also checked an ESR and CRP, which were within normal ranges, 12 mm/h and 6 mg/L, respectively.

Discussion

This study reports a case of MFS, who underwent THA due to severe hip DJD, but unfortunately was complicated with wound dehiscence, surgical site infection (SSI), and development of brachial plexus neuritis post-operatively. To our knowledge, this is the first report of closed-incision negative-pressure wound therapy (ciNPWT) used for the treatment of SSI and wound dehiscence after THA in MFS. It is also the first report of Parsonage-Turner syndrome after THA in MFS.

Despite the defect in the fibrillin-1 gene, MFS patients have normal skin texture and elasticity [12], and poor wound healing is not a typical feature of Marfan, as opposed to Loeys-Dietz syndrome or some types of Ehlers-Danlos syndrome [16]. Although not a common concerning issue, there have been reports of poor wound healing in MFS patients after repair of pectus excavatum, herniorrhaphy, and cleft palate [17-19]. However, no report of poor wound healing in MFS after THA, as in our case, was found in the literature.

Table 1
The patient's four-extremity neurologic examination.

Limb	Muscular group	Motor[a]		Limb segments	Sensory	
		Right	Left		Right	Left
Upper limb	Shoulder abduction	5	4	Arm	Medial side: ↓	Nl
	Elbow flexion	4	4			
	Elbow extension	4	3			
	Wrist extension	5	3	Forearm	Medial side: ↓	Nl
	Finger extension	5	2	Elbow	Medial side: ↓	Lateral margin: ↓
	Finger abduction	5	5			
	Thumb abduction	3	0	Hand	Median territory: ↓↓ Ulnar territory: ↓	Generalized: ↓↓
Lower limb	Hip flexion	4	-[b]	Foot	Nl	Medial margin: ↓
	Hip extension	4	-			
	Knee flexion	4	-			
	Knee extension	4	-			
	Ankle dorsiflexion	4	4			
	Ankle plantar flexion	4	4			
	1st toe dorsiflexion	4	3			

[a] Muscle forces are assessed clinically in a scale of 0-5/5.
[b] The assessment of left hip and knee muscular forces was not accurate due to recent left THA.

Table 2
The immunologic lab profile of the patient.

Lab parameter	Result
ESR (Frequent)	**Positive**
CRP (Frequent)	**Positive**
RF	**3**+
ANA	0.9 (08-1.2: Equivocal)
Anti-dsDNA	**22.4** (>18: **Positive**)
SSB-LA	7.2 (<12: Normal)
SSA-RO	17.3 (12-18: Equivocal)
P-ANCA	7 (<12: Negative)
C-ANCA	4.4 (<12: Negative)
CH50	>90% (90-100%: Normal)
IgA	118 (70-400: Normal)
Serum cryoglobulin	Negative
Anti-cardiolipin IgM	2 (<12: Negative)
Anti-cardiolipin IgG	3.4 (<12: Negative)
Anti-phospholipid IgM	7.9 (<12: Negative)
Anti-phospholipid IgG	4.9 (<12: Negative)
Wright (serum)	Negative
Wright (CSF)	Negative
RPR	Non-Reactive
VDRL (serum)	Non-Reactive
VDRL (CSF)	Non-Reactive
HIV Ab	Negative
HCV Ab	Negative
HBs Ag	Negative
HBc IgM	Negative
HTLV-1 Ab	Negative
HTLV-2 Ab	Negative
HCV Ab	Negative
VZV Ab	Negative
EBV IgM	Negative

Positive values are shown in bold.

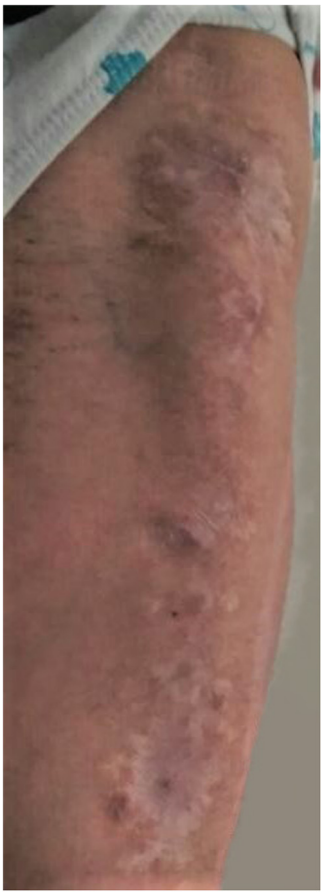

Figure 3. No wound complication at 26-month follow-up.

As mentioned previously, the strongest available evidence regarding NPWT use after arthroplasty emphasizes its prophylactic use in high-risk patients [4-7]. However, its use in treating acute periprosthetic joint infection has been reported only by a few case-series [8-10]. Kirr et al. used NPWT - V.A.C. instill system- after I&D for five cases with acute PJI, three hips and two knees, which resulted in complete wound healing of all cases within 14 days [10]. Lehner and Bernd using the same system for three acute PJI cases, two hips and one knee, could successfully retain all three implants at 8 weeks [9]. Kelm et al. reported the management of 28 acute hip PJIs by I&D followed by internal NPWT, of which 26 had complete eradication of infection at an average of 36 months [8]. In our literature review, no previous report of NPWT use in the management of acute hip PJI in MFS was found. In fact, the studies regarding THA in MFS are rare. The only remarkable study that examined the clinical outcome of THA, that is, the rate and type of complications, in MFS was a retrospective study of 29 Marfan patients (38 hips) from 1977 to 2008. There was a dislocation rate of 10.5% (four hips) at a mean 143.8 ± 35.7 months, all of which required acetabular component reoperation, and a deep infection rate of 5.3% (two hips) at 12 and 44 months. These PJIs were treated by 1-stage and 2-stage total reoperations [20]. However, in our case, the acute dislocation was treated by open reduction surgery, and acute PJI was managed by I&D combined with the more novel ciNPWT.

Although Parsonage-Turner syndrome has been reported after various surgeries [14,15,21], only one report of it after hip arthroplasty was found [22]. Moreover, to our knowledge, this is the first report of PTS in a Marfan patient. In its classic form, PTS is characterized by unilateral shoulder pain and patchy weakness in the brachial plexus distribution [15]. However, the plexopathy could be bilateral in almost 30% of the patients, as in our case [15]. Our patient also had unilateral recurrent laryngeal nerve paralysis and lumbosacral plexopathy. The involvement of nerves outside the brachial plexus has been reported in a minority of PTS cases (2%, recurrent laryngeal nerve; 8.2%, lumbosacral plexus) [15,23].

Summary

Wound dehiscence can occur after THA in Marfan patients. This study has reported the successful management of SSI after THA in a patient with Marfan syndrome. However, as there is strong evidence for prophylactic ciNPWT following THA in high-risk patients, we recommend it in Marfan patients who undergo THA to prevent further complications. Parsonage-Turner syndrome may happen as a rare complication of THA in Marfan. As it could be quite perplexing, the orthopedic surgeons should be familiar with it for the proper diagnosis and management.

Conflicts of interest

The authors declare that there are no conflicts of interest.

For full disclosure statements refer to https://doi.org/10.1016/j.artd.2021.10.006.

Informed patient consent

The author(s) confirm that informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they

have given approval for this information to be published in this case report (series).

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