CASE REPORT

DOI: 10.5336/caserep.2024-104863

Behçet's Disease and Partial Necrosis of Skin Flaps After Breast Reconstruction with Implants

Mehmet Fatih ÖZÇİLER[a], Bilge Kaan İSMAİL[a], Mehmet Sühan AYHAN[a], Osman KURUKAHVECİOĞLU[b]

[a]Gazi University Faculty of Medicine, Department of Plastic, Reconstructive and Aesthetic Surgery, Ankara, Türkiye
[b]Gazi University Faculty of Medicine, Department of General Surgery, Ankara, Türkiye

This case report was presented as a poster at the 45th National Congress of Turkish Society of Plastic, Reconstructive and Aesthetic Surgery, October 28-November 1, 2023, Ankara, Türkiye.

ABSTRACT Behçet's disease is a chronic, relapsing, multisystem disorder characterized by systemic autoimmune vasculitis. It may remain asymptomatic for an extended period or manifest with various complications. The classic triad of Behçet's disease consists of oral ulcers, genital ulcers, and uveitis. Behçet's disease has been associated with rare complications and comorbidities, including mastitis, acral tissue necrosis following minor trauma, severe ulceration of the breast skin envelope, and necrosis of the bilateral nipple-areola complex. Especially; in cases with a positive pathergy phenomenon, trauma-induced triggering of the inflammatory cascade may result in rare cutaneous complications. This case report highlights the association between partial necrosis of bilateral mastectomy skin flaps and Behçet's disease in a 48-year-old female patient who underwent immediate breast reconstruction with direct implant placement following bilateral skin-sparing mastectomy.

Keywords: Behçet's syndrome; breast reconstruction; mammaplasty; autoimmune vasculitis; case report

Behçet's disease is a chronic, relapsing, multisystem disorder characterized by systemic autoimmune vasculitis. It may remain asymptomatic for an prolonged period or manifest with a variety of complications.[1-8] The classic triad of Behçet's disease includes oral ulcers, genital ulcers, and uveitis.[1-7] Blindness, neurological or gastrointestinal system involvement, venous thrombosis, and arterial aneurysms are among the severe symptoms. Additionally, rare complications and comorbidities such as mastitis, acral tissue necrosis following minor trauma, severe ulceration of the breast skin envelope, and necrosis of the bilateral nipple-areola complex have been reported.[3-6,9,11] Notably, in cases exhibiting a positive pathergy reaction, trauma can trigger an inflammatory cascade that may result in rare cutaneous complications.[3,7] Treatment is primarily symptomatic, although corticosteroids or immunosuppressants are required in cases with ocular or neurological involvement.[10]

Given the multisystem nature of Behçet's disease, including widespread vascular inflammation and skin involvement, it is critical to consider this disease when deciding between reconstructive options.

CASE REPORT

A 48-year-old female patient with an 18-year history of Behçet's disease was diagnosed with invasive ductal carcinoma of the right breast through a core biopsy in March 2023 (Figure 1A). She sought simultaneous breast reconstruction following a right side skin-spar-

Correspondence: Mehmet Fatih ÖZÇİLER
Gazi University Faculty of Medicine, Department of Plastic, Reconstructive and Aesthetic Surgery, Ankara, Türkiye
E-mail: drozciler@gmail.com

Peer review under responsibility of Turkiye Klinikleri Journal of Case Reports.

Received: 14 Jul 2024 ***Accepted:*** 10 Oct 2024 ***Available online:*** 07 Nov 2024

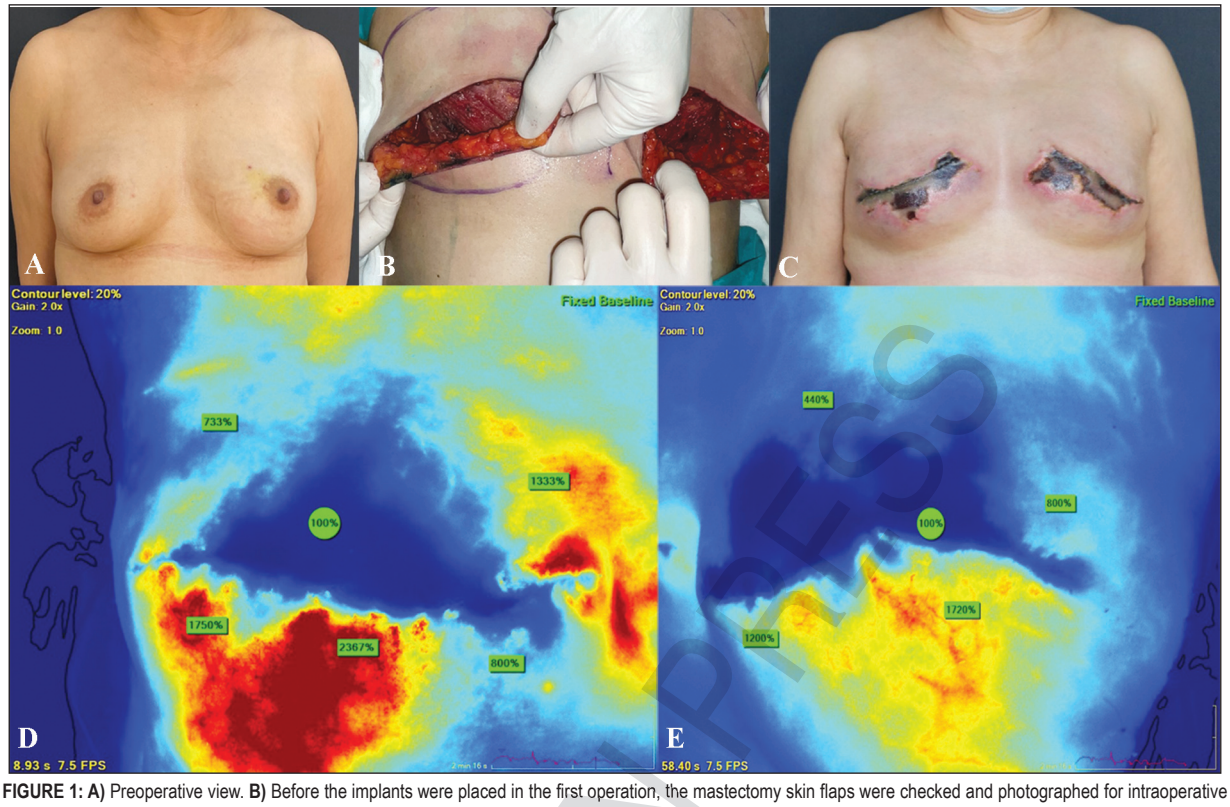

FIGURE 1: A) Preoperative view. **B)** Before the implants were placed in the first operation, the mastectomy skin flaps were checked and photographed for intraoperative thickness and blood supply. **C)** Following the first operation, partial necrosis and demarcation line in the mastectomy skin flaps can be seen. **D)** Intraoperative indocyanine green fluoroscopy of the right breast during surgical debridement and transportation of the implants to the submuscular plane due to the progression of necrosis and demarcation. **E)** Intraoperative indocyanine green fluoroscopy of the left breast during surgical debridement and transportation of the implants to the submuscular plane due to the progression of necrosis and demarcation.

ing mastectomy with sentinel lymph node biopsy and a left side prophylactic skin-sparing mastectomy. According to the information documented in the patient's initial consultation, recurrent oral and genital ulcers, a positive pathergy phenomenon (pustular inflammation secondary to minor trauma), and prior ischemic cerebrovascular disease were observed, characteristic findings of Behçet's disease were noted.

In April 2023, the patient underwent bilateral skin-sparing mastectomies with immediate reconstruction using bilateral 360 cc anatomical implants (Microthane® Polytech Health & Aesthetics GmbH, Germany) placed prepectorally. Intraoperatively, the skin flap thickness and perfusion appeared adequate (Figure 1B). However, during routine postoperative follow-up, the patient-who was not scheduled for chemoradiotherapy and had no history of smoking-developed partial necrosis of the mastectomy skin flaps, particularly around incision margins of both breasts (Figure 1C). Subsequently, surgical debridement was performed at the end of the second postoperative week, and the implants were found to be intact. Although preoperative capillary refill and skin flap color were satisfactory, intraoperative indocyanine green fluorescence angiography revealed localized areas of poor perfusion, inconsistent with clinical findings (Figure 1D, Figure 1E). Following the identification of these areas, the implants were repositioned into the submuscular plane. Before closure, capillary refill was reassessed as neutral, and skin flap color appeared normal, with active dermal bleeding observed.

Despite these interventions, the patient developed signs of infection during subsequent follow-up, and the necrosis progressed, becoming well-demar-

cated. Culture analysis revealed tigecycline sensitive *Acinetobacter baumannii*. Tigecycline therapy was initiated, surgical debridement was performed, and the implants were removed two weeks postoperatively-May, 2023- following the initial debridement. Hematology and rheumatology consultations were requested after routine blood tests revealed thrombocytosis with a progressive increase, likely secondary to the patient's vasculitis. A good response was observed to the combination of surgical debridement and antibiotic therapy. Tigecycline treatment was discontinued on day 14 following infectious disease recommendations. No other complications were reported. Informed consent for publication was obtained from the patient.

DISCUSSION

The pathophysiology of Behçet's disease remains underrecognised. However, histopathological findings typically show vasculitis and thrombosis across all affected organs. Various inflammatory skin lesions are observed in approximately 80% of cases.[1,2] The disease often manifests in the third decade of life and follows a chronic course with relapses and remissions. About 25% of patients experience recurrent superficial or deep migratory thrombophlebitis, potentially leading to vena cava obstruction.[2] The diffuse vasculitis associated with Behçet's disease can cause arterial aneurysms and thrombosis in various regions.[1] In the case presented, widespread vasculitis and inflammatory skin lesions likely contributed to the necrosis of the mastectomy skin flaps. This is supported by the patient's history of ischemic cerebrovascular disease and arthritis.

The pathergy phenomenon is found in most of patient with the diagnosis of Behçet's disease. The positivity of the pathergy indicates cutaneous hyperreactivity to nonspesific minor cutaneous traumas.[13,14] The positive pathergy phenomenon further predisposes patients to the development of severe inflammatory skin lesions.[7-10,12,15]

The bilateral nature of the complications in this case suggests a systemic issue rather than a localized one.[5,6] Although breast involvement in vasculitic pathologies is extremely rare, it can occur as part of a systemic condition or as a localized pathology.[6] The literature describes several cases of breast involvement in vasculitic diseases, most commonly associated with Wegener's granulomatosis and giant cell arteritis.[6] Despite an extensive review of the literature, no reports were found regarding breast reconstruction complications in patients with Behçet's disease.

A rare case of postpartum bilateral nipple-areolar complex necrosis in a 28-year-old female patient with Behçet's disease was reported by Bergant et al.[3] Although skin necrosis secondary to enoxaparin (low molecular weight heparin) was considered in the differential diagnosis, the absence of thrombocytopenia and the patient's tolerance to previous heparin use ruled this out. Similarly, in our case, no complications arose from previous heparin use, and thrombocytosis-rather than thrombocytopenia-was observed, excluding this differential diagnosis.

Ball et al. noted that severe breast ulcers may be considered among the clinical skin manifestations of Behçet's disease.[7] Although spontaneous ulceration was not observed in the presented case, Bulur and Onder emphasized that cutaneous symptoms may be more severe in patients with a positive pathergy reaction, as was seen in this patient.[8]

Özalp et al. reported a case involving a 22-year-old female patient with Behçet's disease who underwent amputation at the mid-diaphyseal level of the proximal phalanx of the left first toe, following an excisional biopsy of a papular lesion on the nail bed.[10] Although angiography of the left lower extremity on the fifth postoperative day demonstrated intact flow in the dorsalis pedis and posterior tibial arteries, with adequate perfusion extending to the digital artery level, a revision amputation at the metatarsophalangeal level was necessary on the twentieth postoperative day due to necrosis progression. Similarly, in the case we present, Doppler ultrasound and angiography indicated patent circulation postoperatively; however, necrosis continued to progress, and the underlying cause could not be clearly identified.

Likewise, according to Bozkurt et al., patients with Behçet's disease who exhibit a positive pathergy phenomenon tend to have a strong inflammatory re-

sponse to minor trauma.[15] Therefore, they recommend avoiding surgical interventions in these patients unless life-threatening conditions.

In our case, an extraordinary complication was observed after a surgical intervention to breasts. From our experience with the presented case, Due to the nature of Behçet's disease, even minor cutaneous trauma can trigger an excessive inflammatory response, which may lead to thrombosis at the vascular level. Therefore, we believe that tissues that appear normal in terms of intraoperative perfusion might undergo acute microvascular occlusion secondary to widespread inflammation-induced thrombosis, ultimately resulting in partial ischemia.

All in all, this case is valuable as it documents a previously unreported complication of Behçet's disease. During the preoperative evaluation of multisystemic inflammatory diseases such as Behçet's disease, it is crucial to conduct a detailed patient assessment and to consider potential risks. Even if the surgical procedure proceeds as planned, these patients may have a higher propensity for complications compared to the general population. As observed in this case, complications such as skin flap necrosis, implant exposure, and infection may arise even when flap thickness is adequate and perfusion appears intact. Our findings, in conjunction with existing literature, suggest that further research should investigate the positive pathergy phenomenon as a potential marker for an increased risk of severe cutaneous involvement following surgery.

In conclusion, the authors recommended a more cautious approach, particularly in patients with positive pathergy tests, and emphasized the importance of thoroughly informing patients about the risk of serious complications. They suggested that considering conservative treatment alternatives might be beneficial for these patients. Also, surgeons should be aware of the development of ischemia in the postoperative period and should inform the patient of the risk.

Additionally, the preoperative history obtained during patient evaluation can provide critical information that directly affects surgical outcomes. The presented case underscores that the surgical process extends beyond the intraoperative period and that the entire perioperative process is directly related to the final outcome.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Mehmet Fatih Özçiler, Mehmet Sühan Ayhan;* ***Design:*** *Mehmet Fatih Özçiler, Bilge Kaan İsmail;* ***Control/Supervision:*** *Mehmet Sühan Ayhan, Osman Kurukahvecioğlu;* ***Data Collection and/or Processing:*** *Mehmet Fatih Özçiler;* ***Analysis and/or Interpretation:*** *Mehmet Fatih Özçiler, Mehmet Sühan Ayhan, Osman Kurukahvecioğlu;* ***Literature Review:*** *Mehmet Fatih Özçiler, Bilge Kaan İsmail;* ***Writing the Article:*** *Mehmet Fatih Özçiler;* ***Critical Review:*** *Mehmet Sühan Ayhan, Osman Kurukahvecioğlu;* ***References and Fundings:*** *Mehmet Sühan Ayhan, Osman Kurukahvecioğlu, Bilge Kaan İsmail;* ***Materials:*** *Mehmet Fatih Özçiler, Bilge Kaan İsmail.*

REFERENCES

1. Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. Lancet. 1990;335(8697):1078-80. PMID: 1970380.
2. Takeno M, Sakane T. Vascular involvement in Behçet's disease. Intern Med. 2001;40(1):3-4. PMID: 11201366.
3. Bergant AM, Widschwendter M, Sepp N. Bilateral nipple ulcers in a breast-feeding woman: a manifestation of Behcet's disease? BJOG. 2000;107(10):1320-2. PMID: 11028591.
4. Konak HE, Erden A, Armağan B, Güven SC, Apaydın H, Dağlı PA, et al. Effects of breast milk on Behçet's disease clinical features. Turk J Med Sci. 2023;53(1):121-9. PMID: 36945965; PMCID: PMC10387996.
5. Soleto MJ, Marcos L. Behçet's disease involving the breast. Eur Radiol. 2002;12 Suppl 3:S98-S100. PMID: 12522614.
6. Dündar SV, Sivri B, Gököz A. Vasculitis of breast in Behçet's disease--a case report. Angiology. 1988;39(10):921-4. PMID: 3177958.
7. Ball E, O'Kane D, McCourt C, McCarron M, Riddell C. Severe breast ulceration in Behçet's treated with cyclophosphamide: a case report. Rheumatology (Oxford). 2021;60(10):e365-e6. PMID: 33895802.
8. Bulur I, Onder M. Behçet disease: new aspects. Clin Dermatol. 2017;35(5):421-34. PMID: 28916023.
9. Hatemi G, Christensen R, Bang D, Bodaghi B, Celik AF, Fortune F, et al. 2018 update of the EULAR recommendations for the management of Behçet's syndrome. Ann Rheum Dis. 2018;77(6):808-18. PMID: 29625968.
10. Özalp B, Akcay C, Aydinol M, Selçuk CT. Toe Amputation after minor surgery in a patient with Behçet's disease: a case report. J Foot Ankle Surg. 2016;55(3):638-41. PMID: 26483163.
11. Goulabchand R, Hafidi A, Van de Perre P, Millet I, Maria ATJ, Morel J, et al. Mastitis in autoimmune diseases: review of the literature, diagnostic pathway, and pathophysiological key players. J Clin Med. 2020;9(4):958. PMID: 32235676; PMCID: PMC7231219.
12. Bhattarai D, Vignesh P, Suri D, Gupta A. Pathergy phenomenon: an important clinical pointer to Behçet disease. BMJ Case Rep. 2019;12(6):e230662. PMID: 31239320; PMCID: PMC6605920.
13. Djawari D, Hornstein OP, Luckner L. Skin window examination according to Rebuck and cutaneous pathergy tests in patients with Behcet's disease. Dermatologica. 1985;170(6):265-70. PMID: 3894082.
14. Gilhar A, Winterstein G, Turani H, Landau J, Etzioni A. Skin hyperreactivity response (pathergy) in Behçet's disease. J Am Acad Dermatol. 1989;21(3 Pt 1):547-52. PMID: 2778116.
15. Bozkurt M, Torin G, Aksakal B, Ataoglu O. Behçet's disease and surgical intervention. Int J Dermatol. 1992;31(8):571-3. PMID: 1428450.