Case Report: Striae- a key clinical sign to look out for when surgically treating patients with Marfan's Syndrome

Authors: Jennifer Huber, DO; Jaya Dhami, MD; Sahejpreet Kaur, MD; Dawn Neil, MD

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Background

Marfan's syndrome (MFS) is a rare hereditary systemic connective tissue disorder due to a mutation in the FBN1 gene in chromosome 15, which produces an extracellular matrix protein known as fibrillin.[1,2] The condition is autosomal dominant, and the prevalence is estimated to be one in 3,000 to 5,000 individuals.[2,3] Marfan's affects many different body parts and can lead to high morbidity and premature mortality.[1] Increased mortality is usually due to aortic root dilatation and dissection. Thus, physicians need to be aware of potential complications when treating patients with Marfan's. In this case report, we present a patient with MFS presenting with dehiscence of the breast post-bilateral reduction mammoplasty.

Literature Review

MFS presents with a broad range of symptoms and severity including rapidly progressive and severe disease involving multiple organ systems in neonates to isolated and minor features of MFS.[2] It is diagnosed according to Ghent nosology, that conducts a comprehensive assessment based on family history and both major and minor manifestations in different organ systems confirming diagnosis in over 95% of individuals.[3,4] The syndrome classically includes cardiovascular, musculoskeletal, and ocular abnormalities, and may also involve the lungs, nervous system, and skin.[2]

Musculoskeletal	Dolichostenomelia (long limbs compared to trunk), Arachnodactyly (abnormally long and thin digits), Thoracolumbar Scoliosis, and Pectus Excavatum and Carinatum, hyperflexible joints
Cardiovascular	Aortic regurgitation, Dilatation, Thoracic Aortic Aneurysm, and Dissection, Mitral Valve prolapse
Ocular	Cataract, myopia, lens dislocation (upward and temporally), retinal detachment
Skin	Striae

Although wound impairment is not a classic sign for Marfan's syndrome, a study done in 2011 suggests that a good diagnostic criterion for MFS includes striae, especially when found in unusual locations. Therefore, MFS has been associated with skin changes and a potential complication to consider with patients with MFS is altered wound healing post-surgery.[5]

Case

A 31-year-old female presented to the plastic surgery clinic for mastopexy vs. breast reduction surgery due to bilateral symptomatic macromastia. She had symptoms of mid-back pain, shoulder discomfort from the strain of bra straps, and striae near the inframammary folds (IMF) and persistent rashes in IMF. She had a pertinent past medical history of Marfan's syndrome and was status-post aortic root and valve repair and abdominal aortic aneurysm repair. Her physical exam was significant for extremely pendulous and dense breasts with grade 3 ptosis, no palpable masses, and fairly symmetric in volume. Patient's height 77 inches, weight 357 lbs, and BMI 42. Measurements of notch to nipple measured 46 cm bilaterally, and nipple to inframammary fold measured 19 and 17 cm bilaterally. The patient underwent bilateral reduction mammoplasty with free nipple grafts. A total of 1934 grams of breast tissue was removed from the left breast and 1747 grams from the right breast, specimens were sent to pathology. Pathology findings included benign breast tissue with autolytic change and suggestion of chronic periductal mastitis. At 1-week post-op, the patient complained of bleeding from incision sites and was found to have minor drainage from the right IMF incision side, key-hole sutures remained intact, and light dressings were applied with recommendations for return to the clinic in 1 week. Follow up at two weeks post-op, the patient had raw areas where the NAC (Nipple-areolar complex) was grafted. After two weeks the patient's NAC was healing with new granulation tissue, but there was a 4 cm area of dehiscence of the right breast exposing breast tissue and a 6 cm opening on the left near the IMF. There was no evidence of hematoma, necrosis, or infection, and the patient was offered vacuum-assisted closure of the wound but declined. Wound cultures grew staph aureus, and the patient was given a course of Doxycycline with plans to return to the OR for revision surgery. The patient underwent a revision procedure for dehiscence of skin about 1-month post-op. The patient tolerated the procedure well and was discharged home. At 4.5 weeks out from debridement, the patient continued to have breakdown of the left IMF which was less severe compared to the first episode of dehiscence. At four months post-op, the patient's left breast healed, right breast continued draining serosanguinous fluid from NAC with plans for a 4 week follow-up. The patient has yet to follow up, currently seven months post-op from debridement.

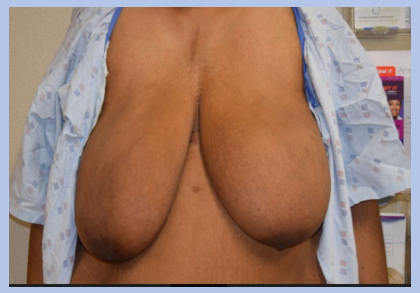

Figure 1- Patient pre-op with extremely pendulous and dense breasts with grade 3 ptosis.

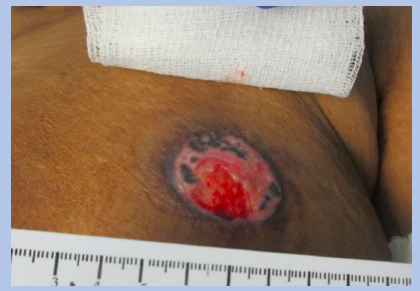

Figure 2- Follow up at two weeks post-op, the patient had raw areas where the NAC (Nipple-areolar complex) was grafted

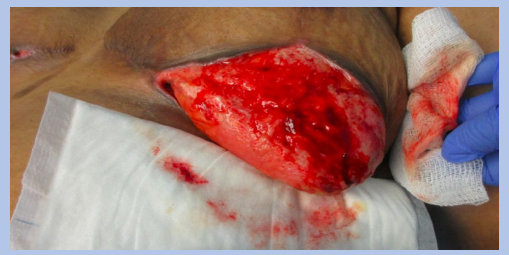

Figure 3- Patient presenting 4 weeks post-op with 6 cm area of dehiscence on the left breast exposing breast tissue near the Inframammary fold.

Discussion

In general, one of the most common complications post-breast reduction is wound dehiscence.[6] As well, the presence of striae in patients with MFS is of diagnostic significance and can lead to delayed wound healing due to abnormal elastic tissue.[7] Our patient suffered severe wound dehiscence at incisions in the IMF and this is aggravated due to the presence of striae. Regardless, diagnosis and treatment of wound dehiscence is the same, with examination of the wound, determination if the wound is infected (ie. wound culture), antibiotics if infection is present, changing wound dressings, open wound to air to speed healing, and negative pressure wound healing. Surgery is required for 1 or more: (1) infected, damaged, and/or dead tissue, (2) add new sutures in wound, (3) mesh placement to help close the wound.[8] Every case is unique and individual and should be managed accordingly.

Contacts:
jennifer.huber@arnothealth.org
jaya.dhami@arnothealth.org