CASE REPORT

Venous Congestion of the Breast Mimicking Inflammatory Breast Cancer: Case Report and Review of Literature

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Abstract: Unilateral breast edema can be worrisome for inflammatory breast cancer. We review the literature and present a clinical case of a patient presenting with features concerning for inflammatory breast cancer, but in fact were the result of dialysis access related venous congestion of the breast.

Key Words: central vein stenosis, nephrogenic systemic fibrosis, unilateral breast edema

Unilateral breast edema can arise from a multitude of etiologies ranging from relatively benign processes such as mastitis or fat necrosis, to the devastating diagnosis of inflammatory breast cancer. Seemingly unrelated, many women with end stage renal disease require upper extremity central vein cannulation for dialysis and many will have had surgically created upper extremity arteriovenous fistulas for long-term dialysis access. The venous stenosis that can arise in the setting of central vein cannulation can be made clinically significant by augmented venous flow associated with an arteriovenous fistula. Depending on the location of the stenosis, this can result in venous hypertension and interstitial edema of the soft tissues. If this occurs distal to the venous drainage of the breast, the resulting congestion can create physical characteristics that mimic those found in inflammatory breast cancer including: unilateral breast edema, erythema, thickened dimpled skin, or peau d’orange appearance. In a patient with unilateral breast edema and a history of previous or current ipsilateral arteriovenous fistulas or central venous cannulation, venography can be very useful in demonstrating a central vein stenosis and engorged collateral vessels which may explain unilateral breast edema. In order to resolve the venous hypertension and breast edema, either balloon angioplasty with or without stent placement, or graft ligation may be necessary. This may obviate the need for an extensive and possibly morbid cancer evaluation.

CASE REPORT

A 29-year-old African-American female with a history of end stage renal disease secondary to congenital dysplasia from Turner syndrome presented initially to an outside facility with marked asymmetry of the breasts. The asymmetry was progressive and developed over the past several months. Her past medical history is otherwise significant for hypertension and a seizure disorder. She has undergone a hysterectomy, parathyroidectomy, cholecystectomy, living-related renal transplantation, and numerous dialysis access procedures including tunneled catheters to the left and right internal jugular veins and subclavian veins, as well as arteriovenous fistulas to the right and left arm veins, first in the forearm and then in the more proximal brachium. Her kidney transplant failed 6 years after it was placed, thereby necessitating a return to hemodialysis therapy. Her most recent and current dialysis access is a left upper extremity arteriovenous graft. She has no family history of malignancy. She does not smoke, drink, or use illicit drugs. She experienced menarche at age 12, is nulliparous, and has never taken hormone replacement therapy.

On initial exam, the patient was thin and demonstrated marked asymmetry of the breasts with the left being significantly larger than the right. There were
obvious skin changes involving the majority of left breast extending up to the level of the clavicle. Her breast skin was thickened and dimpled. She had well healed surgical scars on the upper outer aspect of her nipple areolar complex. There were venous collaterals appreciable across her left chest. There was a left upper extremity arteriovenous graft with a palpable thrill.

She was evaluated with mammography at an outside hospital 2 months prior, which demonstrated an increased density of the left breast. With concern for malignancy in mind, the patient underwent excisional biopsy which showed only dense fibrosis. Further examination at the time of referral was remarkable for persistent breast findings and subtly palpable axillary lymphadenopathy (Fig. 1). Subsequent MRI demonstrated axillary lymphadenopathy and enlargement and thickening of the skin of the left breast without any evidence of an enhancing mass (Fig. 2). The patient then was evaluated with ultrasound guided biopsy of the left axilla which yielded benign lymph node tissue.

With no evidence of a malignant etiology that would account for the breast changes, the patient then underwent venogram of the left sided central veins to rule out a mechanical obstruction in the venous system. Venography revealed a significant stenosis of the left innominate vein (Fig. 3), and multiple balloon angioplasties with a 10 mm balloon produced a moderate reduction in the stenosis (Figs. 4 and 5). There were no other areas of obstruction and the left upper arm arteriovenous loop graft was patent.

The patient experienced dramatic resolution in both the engorgement of the breast and the skin edema; however, this partially recurred within the next 6 months. A subsequent angioplasty of the previous stenosis revealed only partial recurrence of the stenosis which was then successfully ballooned for a second time. The patient has now experienced total resolution of skin symptoms (thickening and pitting) with near
total improvement in breast size. Now 6 months after her second angioplasty, her breasts remain nearly symmetric.

DISCUSSION

Unilateral breast edema can be seen with variety of pathologic processes ranging from congestive heart failure to mastitis (1). Perhaps one the most grave etiologies is inflammatory breast cancer. Inflammatory breast cancer (IBC) is rare, representing only 1–4% of all breast cancers, but more aggressive. More than half of all women diagnosed with IBC will die within 5 years despite receiving optimal therapy (2). As many as 35% of women with IBC have metastatic disease at the time of diagnosis (2). Given the grave nature of IBC, ruling this out of the differential is essential.

Topf et al. (3) were one of the first groups to report on a case of unilateral breast enlargement secondary to central vein occlusion and ipsilateral upper extremity arteriovenous fistula in 1977. Their patient was a 71-year-old woman who presented with unilateral breast edema and was admitted for presumed carcinoma of the involved breast. She was found to have subclavian vein occlusion just proximal to the junction of the mammary and subclavian veins. Ligating her fistula resulted in near complete resolution of her breast symptoms within 1 month.

Pacheco et al. (4) report on a 56-year-old female with progressive enlargement of the right breast after an arteriovenous graft was created in her right forearm. She had previously received dialysis through a right internal jugular vein catheter. Venography demonstrated a right brachiocephalic vein occlusion which was amenable to balloon angioplasty and stent placement. Her breast asymmetry resolved in 2 weeks.

Akan et al. (5) report on a 42-year-old female with unilateral breast enlargement resulting from a dialysis catheter-related subclavian vein stenosis. They hypothesize that impediment of venous return from the mammary vein into the subclavian vein as a result of the subclavian vein stenosis led to the breast asymmetry. They do not comment on how this was ultimately managed, or if any intervention took place.

Gadallah et al. (6) report on a case of marked breast enlargement secondary to an arteriovenous fistula with subclavian vein occlusion proximal to the junction of the mammary vein. Similarly, the diagnosis of inflammatory breast cancer was entertained and excisional biopsy of the breast was performed. Pathologic evaluation revealed fibrosis and duct ectasia without evidence of carcinoma. Later, fistulogram would demonstrate left subclavian vein occlusion proximal to the left mammary vein junction with
filling of multiple vein collaterals. Gadallah noted that in order for breast engorgement to take place the site of the thrombus or stenosis would have to be proximal to the junction of the subclavian vein and mammary vein such that it would be filled by collateral circulation. Furthermore, collateral circulation augmented by the creation of an arteriovenous fistula contributed to the venous hypertension. In this case the breast edema was “well tolerated” and therefore the fistula was not ligated.

Ruiz-Valverde et al. (7) report on unilateral breast and arm enlargement secondary to a hemodialysis arteriovenous fistula without evidence of subclavian vein occlusion. This 61-year-old woman was dialyzed through a left dual chamber jugular catheter and had never had her subclavian vein cannulated. When studied, there was no evidence of stenosis or thrombosis. Upon ligation of her left humeral-cephalic arteriovenous fistula she experienced complete resolution of the breast edema. Ruiz-Valverde hypothesized that venous hypertension alone was enough to result in breast edema. Interestingly, no comment was made regarding the patency of the more proximal central veins, so it is conceivable that there was a stenosis or thrombosis proximal to the patent subclavian vein.

Phillips et al. (8) report on four cases of breast enlargement and central vein stenosis in patients undergoing hemodialysis. Their patients had varying locations of central stenosis and were successfully treated either with PTA or excision of the dialysis graft.

Most recently, Kuerer et al. (9) report on a 74-year-old woman with end stage renal disease who presented with a 2-month history of progressive unilateral breast edema of the left breast and skin changes worrisome for inflammatory breast cancer. Her evaluation included punch and core biopsies which were negative for breast cancer. She had been receiving hemodialysis through left sided internal jugular catheter and also had a left forearm arteriovenous loop graft. Venogram demonstrated left innominate vein stenosis amenable to balloon angioplasty. Her breast edema showed significant improvement over the next few days. Interestingly, this patient had been noted to have elevated venous pressure at dialysis.

Significant subclavian vein stenosis will occur in 50% of patients as early as 2 weeks following placement of a subclavian catheter. Endothelial disruption from catheter insertion as well as trauma induced from the movement of the indwelling catheter, infection, location, and catheter composition have all been suggested as contributing factors to central vein stenosis (10). Phillips et al. (8) point out that the stenosis is often asymptomatic unless the additional venous flow created by an arteriovenous fistula results in venous hypertension and subsequent interstitial edema.

The differential diagnosis for unilateral breast edema in a patient with end stage renal disease also includes a recently described systemic fibrosing disorder known as nephrogenic systemic fibrosis. This poorly understood skin condition is seen in persons with renal insufficiency of various etiologies. There is no standard therapy and the pathogenesis remains unclear. One theory suggests that aberrant wound repair occurs even in the absence of obvious tissue injury. Histologically, there can be a markedly thickened dermis filled with collagen bundles and fibrous bands (11). It is characterized by peau d’orange skin changes and was recently described clinically involving skin changes, tense swelling, and dimpling of the breasts of a 61-year-old woman. This prompted an evaluation for inflammatory breast cancer; however, her biopsy tissue showed an infiltrating pattern of fibroblastic cells consistent with what was formerly known as nephrogenic fibrosing dermopathy.

The breast erythema and thickened dimpled skin, or peau d’orange appearance, seen in cases of central vein stenosis mimics the presentation of inflammatory breast cancer. Considering this diagnosis of central venous obstruction in a patient with unilateral breast edema and a history of upper extremity arteriovenous dialysis fistula may prompt early evaluation with venogram. This may demonstrate a central vein stenosis amenable to angioplasty and thereby obviate the need for an extensive and potentially morbid cancer evaluation. Even a core biopsy or excisional biopsy when undertaken in this setting can be considerably more challenging given the venous hypertension and collateralization. Many of the cases reported in the literature were successfully treated with endovascular therapy including balloon angioplasty and the placement of intraluminal stents. Where appropriate, surgical bypass or ligation of the fistula can be utilized to relieve the obstruction or decrease the flow across an obstruction. These options should generally reserved for those patients with stenoses not amenable to less invasive strategies (10).

Given the paucity of venous access options for our current patient, ligation of her arteriovenous graft is not desirable and will be avoided. Should she
experience a recurrence of symptoms, we will plan to place a stent at the time of angioplasty, acknowledging an increase risk of resultant upstream arteriovenous graft occlusion should the stent occlude. Meanwhile, she is undergoing evaluation by our transplant surgery division for consideration of repeat renal transplant.

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