Case Report

Recurrent and exaggerated secondary vulval lymphoedema with successive pregnancies: An exceptionally rare occurrence

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Abstract

Lymphoedema is swelling due to the excess accumulation of lymph in the tissues caused by inadequate lymph drainage owing to primary or a secondary etiology. Here we present a rare case of secondary lymphoedema of the vulva which exaggerated with each successive pregnancy with considerable remission in between with residual mass left behind following termination of each pregnancy, though the size of the remnant mass increased at the end of each successive pregnancy.

Keywords: Vulval mass, recurrent lymphoedema, secondary lymphoedema.

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INTRODUCTION

Lymphoedema is swelling due to the excess accumulation of lymph in the tissues caused by inadequate lymph drainage. Primary lymphoedema implies a genetic or constitutive cause whereby there is an intrinsic fault in lymph drainage determined by a lymphatic maldevelopment or functional weakness. Secondary lymphoedema implies an acquired failure of previously normal lymph drainage due to an identifiable cause extrinsic to the lymphatic system.[1] Repetitive exaggeration of secondary vulval lymphoedema with each subsequent pregnancy, with a residual component of lymphoedema left behind following gradual and spontaneous regression after the termination of each pregnancy, is an exceptionally rare case to be witnessed, which is discussed below.

CASE REPORT

A 32 year old female, gravida 5, para 4, living 4, was referred from a peripheral hospital at nine months of gestation with a huge mass in the perineum. On eliciting patient's history, it was found that the patient first noticed a small vulval mass during her first pregnancy, 10 years back at around 6-7 months of gestation, which gradually progressed to a size of lemon by term, followed by a gradual regression in size following delivery, only to leave a small residual mass. The patient observed a gradual increase in the size of the mass again, more apparent at around 6-7 months of gestation, during her subsequent pregnancies, with a progressive increase in the total size, with each pregnancy. And following delivery, there was again regression in size every time, but the residual mass size increased with each successive pregnancy. Surprisingly, patient had a spontaneous vaginal delivery every time, without any need for intervention. In the present pregnancy apart from the mass, patient had frequency and urgency of urination starting from 7 months of gestation. There was also no family history suggestive of lymphedema. On examination, patient was thin built,pale, with a term sized

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uterus, with a huge perineal mass having a horse-shoe shaped origin involving mons pubis, clitoris and vulva on either side, stretching the urethral opening. The mass was huge enough to hang up to the level, slightly above the knees in standing position, with irregular surface, studded with multiple polyp like / papillary projections of varying size, with oozing of foul smelling whitish fluid from few of them. There were also bilaterally symmetrical inguinal incision scar marks noted, which the patient related to surgery at the age of 5 years, when she was diagnosed with tuberculosis. However, there were no records regarding surgery or tuberculosis that could be recovered from the patient.

Figure 1

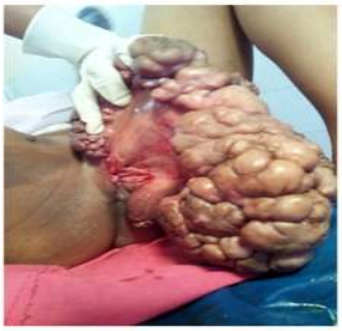

Figure 2

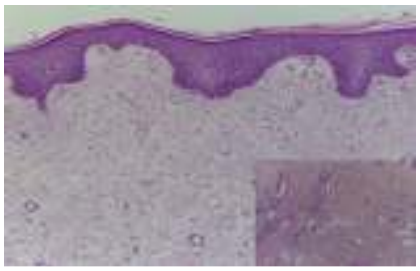

Figure 3

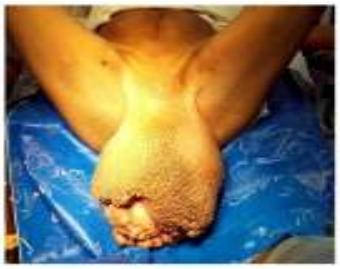

Figure 4

Figure 5

Figure 6

Figure 7

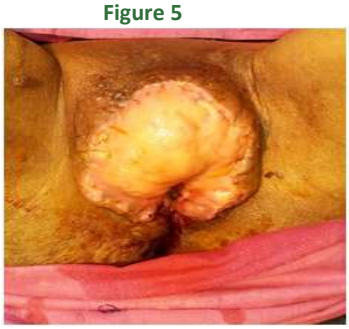

Figure 8

Legend

Figure 1: Vulval mass accompanying pregnancy at term

Figure 2: Picture showing distribution of the origin of mass and stretching of the urethra

Figure 3: Biopsy from the vulval mass showing focal atrophy of the lining squamous epithelium with edema and perivascular collections of lymphocytes and plasma cells in the subepithelial tissues (HandE, 100x). Deeper tissues showing collections of dilated lymphatics (Inset).

Figure 4: Picture of the mass as seen on day 10 post LSCS

Figure 5: post operative specimen

Figure 6: Huge perineal mass weighing 2800gm

Figure 7: Reconstructed vulva at day 1 vulvectomy

Figure 8: Reconstructed vulva on day 10 postvulvectomy

On investigation, haemoglobin was found to be 6.1g%.Ultrasonography of gravid uterus revealed a live fetus of term gestation. USG of the mass revealed heterogeneously hyperechoic soft tissue with multiple vessels traversing through it. However, only superficial part of the mass could be visualised on Ultrasound. Peripheral blood smear was negative for filariasis. RtPCR of the specimen from mass was negative for TB bacilli. Histopathological examination of the tissue from the mass showed squamous epithelial lining with focal atrophy,

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with subepithelial oedema and perivascular collections of lymphocytes and plasma cells without koilocytic or dysplastic changes, compatible with vulval lymphedema (Figure 3) Deeper tissues showed collections of dilated lymphatics (Inset). Patient was transfused 2 units of blood. Patient went into labour spontaneously. There was difficulty encountered to introduce a catheter and to do a per vaginal examination. Patient was taken up for emergency LSCS as it was considered to be difficult or impossible to deliver vaginally because of the growth. A healthy male baby of 3.2 kg was delivered and intraoperatively there was no mass or abnormality identified. On day 10 puerperium, vulvectomy was done and the huge perineal mass weighing 2800gm (Figure 6) was excised and sent for HPE and the report was consistent with that of the earlier sampling. The reconstructed vulva (Figure 7) was slightly oedematous with mild oozing of lymph like fluid. Catheter was kept in situ for 5 days. Absorbent dressing was done for 5 days, followed by removal of the sutures on the 10th post operative day. (Figure 8)

DISCUSSION

The pathological processes involved in secondary lymphoedema following acquired obstruction or obliteration of lymph-conducting pathways include infection, inflammation, trauma (including surgery and radiation) and malignant disease. Trauma to lymphatics, usually needs to be extensive to induce lymphoedema. Failure of lymphatics to regenerate and re-anastomose satisfactorily through scarred tissue is probably responsible for lymphoedema.[1]

Decreased transport of lymph from the skin leads to an increase in protein-rich interstitial fluid. This rises the interstitial pressure consequently dilating the lymphatics.[1]

Temporal changes observed in experimental lymphoedema indicate that the collagen fibres initially become swollen and separated. Mononuclear cells are seen around the lymphatic and blood vessels. Lymphatic walls thicken and fibrose. The muscular elements of the collecting trunks atrophy. Macrophages, fibroblasts and lymphocytes accumulate. Overgrowth of the interstitial connective tissue gradually transforms the soft stage of lymphoedema into the hard late-stage form. The simple excess of protein seems to be the cause of the fibrosis. The number of blood vessels also greatly increase.[1]

It is said that no form of lymphoedema is mutually exclusive, and frequently a number of factors combine to produce swelling. For example, lymphoedema may become clinically obvious only when the lymphatic load is increased through higher fluid filtration, exhausting lymph drainage capacity. However, the puzzle still lies unveiled, as to why there can be such a delay up to 20 years before lymphoedema manifests.[1]

This delay in manifestation is evident in our case too, where the patient developed lymphoedema after almost around 17 years of lymphatic affection. Patient's history suggestive of tuberculous lymphadenitis being diagnosed in her childhood, accompanied by the bilateral scars in the inguinal region, helps to label this case to be one of the secondary lymphoedemas, which has become evident due to increased lymphatic load owing to pregnancy.

CONCLUSION

Lymphedema associated with pregnancy is a rare occurrence to be witnessed. Recurrent lymphedema with successive exaggeration of clinical presentation is an even rarer presentation.

REFERENCES

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