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Pseudosarcoma – massive localized lymphoedema in morbidly obese – a rare entity: Case report

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ABSTRACT

INTRODUCTION: Massive localized lymphoedema (MLL) first described in 1998 by Farshid and Weiss. Usually MLL present like huge pedunculated mass and appear like sarcoma hence called Pseudosarcoma. Morbid obesity is a growing epidemic in our society. Morbid obesity is usually associated with hypertension, Diabetes mellitus, dermatological complications like Acanthosis nigricans, skin tags, leg ulcers, edema, lymphoedema, plantar hyperkeratosis and massive localized lymphoedema (MLL) is one of the complications of morbid obesity.

Pseudosarcoma is due to derangement of lymphatic channels secondary to excessive deposition of adipose tissue.

PRESENTATION OF CASE: We report a patient afflicted with this unique disorder presented with huge mass arising from mons pubis in morbidly obese individual with body mass index (BMI) 55.

DISCUSSION: Massive localized lymphoedema presenting like pseudosarcoma in morbidly obese individuals is rare. Awareness of this disease is essential to avoid misdiagnosis as soft tissue neoplasm. It is a term used to describe a benign over growth of lymphoproliferative tissue in morbidly obese patients. Because of its size patients have difficult to do daily activities. Histopathologically characterized by dilated lymphatic channels with fibrotic and edematous tissue, without evidence of malignancy. Patient seeks treatment only if there is huge swelling causing discomfort, complications like excoriation, wound break down occur. The treatment of choice is complete excision.

CONCLUSION: Surgical treatment is effective if done along with bariatric surgery. Functional rehabilitation was achieved. No recurrence was observed within the follow up period of twenty months and BMI was reduced to 28.

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1. Objective

Massive localized lymphoedema presenting like pseudosarcoma in morbidly obese individuals is rare. Awareness of this disease is essential to avoid misdiagnosis as soft tissue neoplasm. It is a term used to describe a benign over growth of lymphoproliferative tissue in morbidly obese patients. Clinically pseudosarcoma presents like a huge mass most commonly arising from lower extremities. Awareness of this entity, clinical correlation and gross pathological correlation are essential in the separation of this distinctive pseudosarcoma from its various morphological mimics. Surgical treatment is effective if done along with bariatric surgery and functional rehabilitation was achieved.

2. Introduction

Morbid obesity is a growing epidemic in our society. Morbid obesity is usually associated with hypertension, Diabetes mellitus, dermatological complications like Acanthosis nigricans, skin tags, leg ulcers, edema, lymphoedema, plantar hyperkeratosis and massive localized lymphoedema (MLL) is one of the complications of morbid obesity. MLL first described in 1998 by Farshid and Weiss.¹² Usually MLL present like huge pedunculated mass and appear like sarcoma hence called Pseudosarcoma. Pseudosarcoma is due to derangement of lymphatic channels secondary to excessive deposition of adipose tissue. Pseudo sarcomas are composed of fibroblasts and myofibroblast includes nodular fasciitis, proliferative fasciitis, myositis, intravascular fasciitis etc. Lesions shows striking smooth muscle hyperplasia. Lack of staining by antibodies against murine double minute 2protein and cyclin dependent kinase4 and absence of Group AT hook2 transcription factor rearrangement by fluorescence in situ hybridization support diagnosis of MLL. Treatment of choice is complete surgical excision. Pseudosarcoma is not a malignant one, but local recurrence may occur in 14–50% of cases. Complications of pseudotumor include skin excoriation, difficult daily activities & rarely malignant transformation

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Fig. 1. Pre operative view of the swelling.

to liposarcoma, angiosarcoma. We report a case of a patient presented with a massive benign soft tissue tumor of mons pubis,⁶ which has been named as pseudo sarcoma in the literature.

3. Presentation of case

A female patient of age 58 years came with known history of hypertension, hypothyroidism, obesity (150 kg – weight). Came with a chief complaint of a huge swelling present at lower abdominal area since 6 years.

Physical findings: Huge mass present between the thighs, which is arising from mons pubis. Patient was morbidly obese with BMI – 55 (Fig. 1).

Investigations: CECT abdomen: no evidence of significant abnormality detected/no evidence of hernia.

Treatment: complete excision of the tumor along with Laparoscopic sleeve gastrectomy for morbid obesity done under general anesthesia.

Macroscopic appearance: fatty fragments of tissue 2 cm × 2 cm × 1 cm to 2.5 cm × 2 cm.

Histopathological examination: pseudosarcoma specimen section revealed fibrofatty tissue – free from malignancy (Fig. 2).

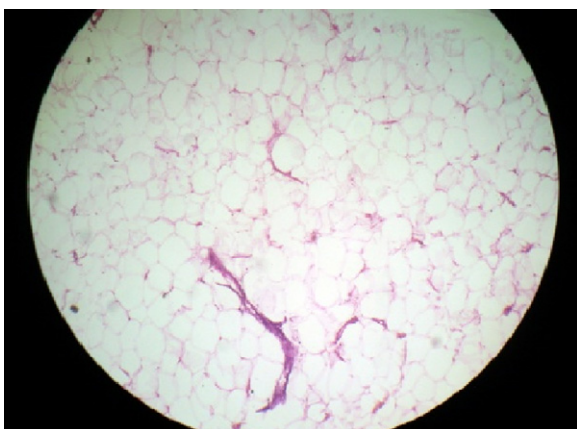


Fig. 2. Pseudosarcoma – microscopic appearance (pseudosarcoma specimen section revealed fibrofatty tissue – free from malignancy).



Fig. 3. Post operative view of surgical site.

Post operative period uneventful. Patient discharged on 5th post operative day.

Follow up of the patient done for twenty months – now the weight of the patient was 70 kg with BMI – 28 (Fig. 3).

4. Discussion

Massive localized lymphoedema is a rare disease in morbidly obese individuals. Only few cases have been reported in the literature.^{1–5} Literature reviewed only 41 cases. MLL is an emerging complication of obesity epidemic, caused by obstruction of lymphatics. MLL present as a giant swelling. MLL is also called as pseudo sarcoma because of its morphological and pathological similarity to sarcoma. Histologically striking dermal fibrosis, expansion of fibrous septa between fat lobules with increased numbers of stromal fibroblasts, lymphatic proliferation and lymphangiectasia, multinucleated fibroblastic cells, marked vascular proliferation present. Moderate stromal cellularity and fascicular growth raised concern among referring pathologist for atypical lipomatous tumor – liposarcoma,⁸ angiosarcoma, fibromatosis.

Other varieties of pseudotumors are nodular fasciitis, intravascular fasciitis, Proliferative fasciitis myositis, cranial fasciitis, Atypical

decubital fibroplasias, postoperative spindle cell tumor, Bizarre parosteal osteochondromatous proliferation, inflammatory pseudotumor, Myositis ossificans, florid reactive periostitis, subungual exostosis have different etiological causes like trauma. But massive localized lymphoedema causing pseudosarcoma seen only in morbidly obese individuals which is due to obstruction of lymphatics. As massive localized lymphoedema occurs due to compression/obstruction of lymphatics by heavy folds of dependent fat. Ferrell proposed that secondary lymphedema development whether due to morbid obesity, cancer, trauma, or heredity factor might be controlled by the same set of genes. Therefore, it is reasonable to assume that genetic factors at least partially control the development of lymphedema in persons who are obese.

Although no studies directly examine the relationship between obesity and the development of lymph edema, indirect evidence from different sources suggests a relationship exists. Evidence from animal models also suggests that impaired lymph drainage, stress, and inflammatory disorders are factors involved in selective hypertrophy of lymphoid tissue-associated adipose tissue. An animal study that investigated the effect of the prox1 master gene haploid condition in mice (i.e., prox1+/-) suggests that minor defects in lymphatic system development, such as the leakage of lymph from abnormally formed lymph vessels, stimulates adipocyte development (preadipocyte differentiation), which in turn promotes adult-onset obesity through site-specific fat deposition. This finding has been corroborated using transgenic mice with different defects in lymph angiogenesis. Together, these studies infer that the development of secondary lymph edema is not just the result of lymphatic dysfunction but also might be linked to lipid metabolism with inflammation as a mediator. Further, under certain conditions, lymph-containing factors may stimulate fat deposition, which can lead to obesity. In a prospective imaging study of the functional status of epifascial and subfascial lymphatic compartments using two-compartment lymphoscintigraphy, Brautigam et al. noted that in edema due to central venous insufficiency a high-volume lymphatic overload was present in the epifascial compartment. However, the same investigators did not see any change in lymph transport in cases of lipedema. This is not necessarily the case; the effect on the lymphatic system seems to depend on the severity and progression of lipedema and whether other co morbidities are present. In any case, the crucial factor is the mechanism by which fatty deposits affect lymphatic function and vice versa. These questions have yet to be resolved.

Although resection is usually successful, recurrence of the problem is less if excision of the tumor done along with bariatric surgery as there is vice versa relationship between the morbid obesity and the lymphoedema.

These tumors grow very slowly. Patient tend to seek treatment only when they reach a particular size to alter their activities of daily living, excoriation, wound break down (or) suspicious of malignant transformation. Therapy of choice is complete surgical excision.

Although recurrence is common, overall prognosis is good. In the present case excision of the mass arising from the mons pubis done along with laparoscopic sleeve gastrectomy. Only few cases are reported in the literature and a good clinical and pathological correlation required to rule out malignancy. During follow up period of twenty months patient achieved good physical rehabilitation without recurrence and BMI was reduced to 28.

5. Conclusion

Surgical treatment is effective if done along with bariatric surgery – Sleeve gastrectomy. Functional rehabilitation was achieved. No recurrence was observed within the follow up period of twenty months and BMI was reduced to 28. The diagnosis of MLL was continues to be challenging, in particular for pathologist. Awareness of this entity, clinical correlation and gross pathological correlation are essential in the separation of this distinctive pseudosarcoma from its various morphological mimics. Within the follow up period overall prognosis was good and no recurrence was found.

Conflict of interest

None declared.

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Author contributions

Data analysis, study design Dr. T. Narayana Rao. Data collection, writing by Dr. Suvarchala Akkidas/Dr. G. Krishna babu.

References

- Goshtitasby P, Dawson J, Agarwal N. Massive localized lymphoedema of the morbidly obese. *Obesity surgery* 2006;**16**:88–93.
- Manduch M, Oliveira AM, Nascimento AG, Folpe AL. Massive localised lymphoedema: a clinicopathological study of 22 cases and review of the literature. *Journal of Clinical Pathology* 2009;**62**:808–11.
- Weston S, Clay CD. Unusual case of lymph edema in a morbidly obese patient. *Australian Journal of Dermatology* 2007;**48**(2):115–9.
- Crystal DJ, O'loughlin B. *Massive localized lymphedema of the thigh*. Queensland, Australia: Department of surgery, Royal Brisbane hospital; 2007.
- Jensen V, Mitwittle, Latifi R. *Massive localized lymphoedema pseudosarcoma in a morbidly obese patient*. Tucson, AZ, USA: Department of Surgery, University of Arizona College of Medicine; 2006.
- Brewer M, Singh DP. *Massive localized lymph edema: review of an emerging problem and report of a complex case in the mons pubis*. Baltimore, MD: Division of Plastic Surgery, Department of Surgery, University of Maryland Medical Center; 2011 May.
- MLL in the morbidly obese, a histologically distinct reactive lesion simulating liposarcoma. *American Journal of Surgical Pathology* 1998;22.
- Farshid G, Weiss SW. MLL in the morbid obese; ahistologically distinct reactive lesion simulating liposarcoma. *American Journal of Surgical Pathology* 1998.

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