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LYMPHOMATOID PAPULOSIS MANIFESTING AS A BLEEDING BREAST NODULE: AN UNUSUAL CLINICAL PRESENTATION.

Breast Surgery

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ABSTRACT

Introduction and Importance Lymphomatoid papulosis is an extremely rare dermatological condition, which falls in the category of lymphoma. The word itself means a lesion which is self-healing and has a pattern of rhythmical paradoxical eruption. It is based on the histopathological diagnosis and a clinical diagnosis of it is challenging. **Case Presentation** We describe a case of a 17-year-old girl presenting with a bleeding nodule from her left breast. It was a 5x4 cm red, fleshy, raised lesion over the medial aspect of the left breast with firm consistency without any tenderness. On performing the ultrasound it showed that it was a superficial lesion. Due to continuous bleeding in spite of compression, it was excised due to cosmetic and symptomatic reasons. Post excision the histopathology report was suggestive of a lymphomatoid papulosis. Though multiple treatment modalities are described for this lesion, excision was appropriate, as we could not wait due to the bleeding. **Clinical Discussion:** Lymphomatoid Papulosis is a very rare condition and has never been described as a de novo lesion in the breast. It has different histological variants and different treatment options. According to the literature review, it is better to have non-operative management as it is a condition that has a high tendency to recur. Options range from observation, phototherapy to biological agents and surgical excision. However, a lesion in an area with cosmetic importance which is bleeding has not been described. Hence, a surgical excision is what we offered to the patients and the condition has been under control. **Conclusion:** Hence, we conclude that, since our patient was a young girl and bleeding from it, we excised it with a 1 cm margin as indicated cosmetically and for a symptomatic cure.

KEYWORDS

Lymphomatoid Papulosis Breast Bleeding Excision

INTRODUCTION

Lymphomatoid papulosis is a rare dermatologic condition which develops as a papulo-nodular lesion. "Lymphamatoid papulosis " literally means a lesion which is self-healing and has a pattern of rhythmical paradoxical eruption. It is a non-aggressive lymphoma but can have histological features similar to malignant lymphoma. It is typically seen as a recurrent crop of papules, which are pruritic at multiple stages of development.

The aetiology of this group of diseases has not been established as yet but viral precursors have been studied. (1) It has more predisposition in middled population and less in extremities of age while,it sis more common in women than men. (2)These lesions heal over a couple of months and leave a depressed scar (3). It is sometimes included in the classification of cutaneous anaplastic large cell lymphoma, in the group of T-cell proliferations expressing CD 30 (4). There are multiple treatment options ranging from conservative measures, medical therapy, cytotoxic therapy, and phototherapy to surgical excision. It is usually preferred to have a non surgical treatment option when the lesion exists in parts of the body like the trunk or limbs as it a condition which has high possibility of recurrence. Surgical excision is rarely done in such cases. However, in our case it was required for a cosmetic as well as therapeutic cure for the patient. This is a very rare condition hence, despite having a wide variety of treatment options there is no established standard of care. In order to reduce the chances of recurrence in such cases, we recommend excision of the lesion with margins of 1cm.

Case Presentation

A 17-Year-old girl presented to the outpatient department, with complaints of bleeding from a raised lesion over her left breast, which had appeared before 1 month. The bleeding had started before 2 to 3 days. She had no complaints like pain, itching, or any palpable lump or nipple discharge with it. There were no systemic symptoms like fever, weight loss or appetite changes. Examination revealed a 5x4 cm red, fleshy, raised lesion over the medial aspect of the left breast, at 10: 00 position. It was 10 cm away from the nipple areola complex as shown in FIGURE 1. The lesion had firm consistency without any tenderness. There were no other lumps in the same breast or axilla. The opposite breast and axilla were normal. There was a small ulcer over the lesion with oozing blood. It had started bleeding spontaneously and conservative treatment such as ice packs and local compression were given. However, they failed to stop the bleeding. Ultrasound of the breast revealed a lesion arising from the dermis without any invasion into surrounding structures. The lesion continued to bleed and hence was offered excision biopsy with wide margins of 1 cm. The excised specimen is shown in Figure 2. The postoperative course was uneventful, and the patient was discharged on the same evening. The histopathology report turned out to be a lymphomatoid papulosis. It showed moderately dense superficial and deep perivascular and peri-appendiceal infiltrate of small and large lymphocytes with papillary dermal oedema. The overlying epidermis showed mild focal spongiosis and slight infiltration of small and large lymphocytes. The CT scan of the chest and abdomen was done to rule out asymptomatic lymphoma and did not reveal any abnormality.

DISCUSSION

In 1968, Macaulay coined the term "lymphomatoid papulosis" when he was describing “a self-healing rhythmical paradoxical eruption, histologically malignant but clinically benign" (5). It was initially considered a pseuodolymphomatous inflammatory process because of its waxing and waning clinical course. It accounts for about 12% of cutaneous lymphomas. The mean age of onset varies between 35 and 45 years. The male-to-female ratio is 1: 5. The aetiology of lymphomatoid papulosis is unknown. A viral aetiology has been suggested. There are multiple reasons for its spontaneous regression, which have not been clearly explained. It is possibly due to the apoptosis of the neoplastic T cells due to the interactions between CD 30 and its ligand.

There have been a few cases of lymphomatoid papulosis reported in the literature as case reports and case series. Most of them developed in the area of prior treatment or over an existing disease. Very few cases, as in our case report have described a de- Novo lesion. J J Scarisbrick et al described regional lymphomatoid papulosis in four patients, in areas of flank and buttock. Amongst the four one patient had mycosis fungoides, and the lymphomatoid papulosis was confined to the same area where the patient had mycosis fungoides (6) Rosario Haro et al reported a case which had lymphomatoid papulosis in the beast in the region of radiation which was given after partial mastectomy as a treatment of invasive ductal carcinoma. They described this phenomenon as “recall dermatitis” as it developed after radiation therapy.(7)

According to the number of lymphocytes expressing CD30, and according to their size, it is classified into three types (8). Type A papulosis, which has multiple CD 30 on the cells. Type B is similar to mycosis fungoides with very few or nearly no CD 30. And type C has large CD30+ lymphocyte plaques. A few reports have an extended classification of Type D and Type E. Werner Kempf et al described Angio invasive lymphomatoid papulosis which they termed as Type E. Type E is characterised by oligolesional papules with angiocentric and angio destructive infiltrate of lymphocytes with CD 30. Clinically they tend to develop ulcers which ultimately develop large necrotic eschars. (9) There is Type F as well, which is not included in the WHO classification. (10) These will include patterns with perifollicular

infiltrates and fol- liculotropism. A new genetic variant LyP with chromosome 6p25 rearrangement has also been recognized by the WHO classification 2016. (11)

There are multiple treatment options irrespective of histological subtypes. Lymphomatoid papulosis is a highly recurrent disease and hence conservative treatment options are usually preferred. (12) Methotrexate can be given orally, subcutaneously, or intramuscularly. It is started as a low-dose regimen ranging from 25- 30 mg per week. It is usually continued up to 4-8 weeks. Once the disease is controlled in terms of reduction of the number and size of lesions along with symptomatic relief, the dose is gradually tapered to the lowest possible dose. (13) Folic acid is usually given with methotrexate to reduce the side effects of methotrexate like hepatic fibrosis, gastrointestinal symptoms, mouth ulcers and cytopenias.(14) The second option is phototherapy. PUVA therapy has been proposed to treat lymphomatoid papulosis. It is said that the lesion would disappear after about 12- 15 sessions of this therapy, but recurrences do happen. ESMO publishes guidelines for primary cutaneous lymphomas in 2018. They described local radiation as a treatment option with a dosage of 20-24 Gy with 2 cm margins. (15)Vidhatha Reddy et al have described the use of excimer laser for the treatment of lymphomatoid papulosis which presented as a lesion on the hand of a patient. It worked on the principle of DNA breakage. It did not prove to be curative but gave symptomatic relief to the patient.(16) The local chemotherapies (solution or gel of mechlorethamine), bexarotene, and interferon is sometimes used as therapeutic alternatives. If there are lesions, surgical excision or radiotherapy can be proposed if spontaneous resolution does not occur after a period of 4 to 12 weeks (17) (18). Surgical excision has also been described as a treatment option if the lesion does not resolve with local or systemic therapies. Though a few cases show high rates of recurrence. Kemp f et al used excision as a primary mode of treatment in 19%-57% of the patients. And they reported relapse in 43% of cases. (19) A study done at Stanford done by Howard Liu et al stated that either surgical excision or conservative management had similar survival rates with over all survival of 92% at 5 and 10 years. (20) . There is no consensus about the ideal surgical margins when excision is attempted. In our case, we were compelled to excise the lesion due to its bleeding and location. The surgical margin kept was 1 cm and the lesion has not recurred after a 2-year follow-up period. So, we can conclude that a margin of 1 cm should be adequate. Hence, there is no survival advantage of surgical excision if the lesion is benign but in our case, the lesion was actively bleeding hence excision with adequate margins provided a symptomatic as well as a cosmetic cure for the patient.

CONCLUSION

Lymphomatoid papulosis can present as a bleeding mammary nodule and we recommend performing surgical excision with margins as wide as 1 cm to provide therapeutic cure, cosmetic outcomes as well as to rule to malignancy.

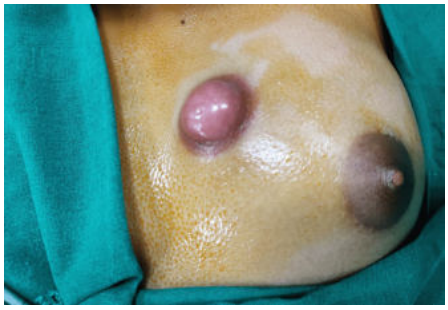

Figure 1: Pre-operative Clinical Photograph of the lesion

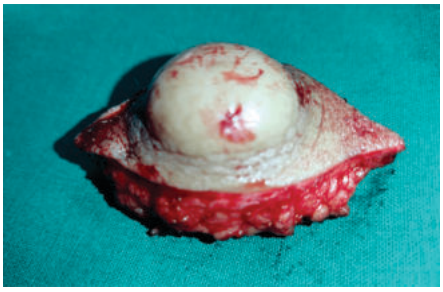

Figure 2: Excised Specimen with 1 cm margin

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Consent Written

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Credit Authorship Contribution Statement

All authors equally contributed to the analysis and writing of the manuscript.

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REFERENCES

1. Toumi A, Fazal S, Litaiem N. Lymphomatoid Papulosis. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 [cited 2020 Oct 23]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK532295/
2. Wagner G, Rose C, Klapper W, Sachse MM. Lymphomatoid papulosis. JDDG J Dtsch Dermatol Ges [Internet]. 2020 [cited 2020 Oct 23];18(3):199–205. Available from: https://onlinelibrary.wiley.com/doi/abs/10.1111/ddg.14041
3. Lymphomatoid papulosis | Genetic and Rare Diseases Information Center (GARD) – an NCATS Program [Internet]. [cited 2020 Oct 23]. Available from: https://rarediseases.info.nih.gov/diseases/6944/lymphomatoid-papulosis
4. Sauder MB, O'Malley JT, LeBoeuf NR. CD30+ Lymphoproliferative Disorders of the Skin. Hematol Oncol Clin North Am [Internet]. 2017 Apr [cited 2020 Oct 23];31(2):317–34. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5776746/
5. Haserick JR. Lymphomatoid papulosis Macaulay's disease. Am J Dermatopathol [Internet]. 1981 Jul [cited 2020 Oct 23];3(2):177–8. Available from: https://journals.lww.com/amjdermatopathology/citation/1981/00320/lymphomatoid_papulosis_macaulay_s_disease.8.aspx
6. Scarisbrick JJ, Evans AV, Woolford AJ, Black MM, Russell-Jones R. Regional lymphomatoid papulosis: a report of four cases. Br J Dermatol. 1999 Dec;141(6):1125–8.
7. Haro R, Juarez A, Díaz JL, Santonja C, Manzarbeitia F, Requena L. Regional Lymphomatoid Papulosis of the Breast Restricted to an Area of Prior Radiotherapy. :5.
8. Kempf W, Mitteldorf C, Karai LJ, Robson A. Lymphomatoid papulosis - making sense of the alphabet soup: a proposal to simplify terminology. J Dtsch Dermatol Ges J Ger Soc Dermatol JDDG. 2017 Apr;15(4):390–4.
9. Kempf W, Kazakov DV, Schärer L, Rütten A, Mentzel T, Paredes BE, et al. Angioinvasive lymphomatoid papulosis: a new variant simulating aggressive lymphomas. Am J Surg Pathol. 2013 Jan;37(1):1–13.
10. Pierard GE, Ackerman AB, Lapiere CM. Follicular lymphomatoid papulosis. Am J Dermatopathol. 1980;2(2):173–80.
11. Karai LJ, Kadin ME, Hsi ED, Sluzevich JC, Ketterling RP, Knudson RA, et al. Chromosomal rearrangements of 6p25.3 define a new subtype of lymphomatoid papulosis. Am J Surg Pathol. 2013 Aug;37(8):1173–81.
12. Vergier B, Beylot-Barry M, Pulford K, Michel P, Bosq J, de Muret A, et al. Statistical evaluation of diagnostic and prognostic features of CD30+ cutaneous lymphoproliferative disorders: a clinicopathologic study of 65 cases. Am J Surg Pathol. 1998 Oct;22(10):1192–202.
13. Vonderheid EC, Sajjadian A, Kadin ME. Methotrexate is effective therapy for lymphomatoid papulosis and other primary cutaneous CD30-positive lymphoproliferative disorders. J Am Acad Dermatol. 1996 Mar;34(3):470–81.
14. Shea B, Swinden MV, Tanjong Ghogomu E, Ortiz Z, Katchamart W, Rader T, et al. Folic acid and folinic acid for reducing side effects in patients receiving methotrexate for rheumatoid arthritis. Cochrane Database Syst Rev [Internet]. 2013 May 31 [cited 2022 Aug 12];2013(5):CD000951. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7046011/
15. Willemze R, Hodak E, Zinzani PL, Specht L, Ladetto M. Primary cutaneous lymphomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. Ann Oncol [Internet]. 2018 Oct [cited 2022 Aug 12];29:iv30–40. Available from: https://linkinghub.elsevier.com/retrieve/pii/S092375341931693X
16. Reddy V, Thibodeaux Q, Bhutani T, Sugarman JL. Lymphomatoid papulosis successfully managed with excimer laser maintenance therapy. JAAD Case Rep [Internet]. 2020 Jun 1 [cited 2022 Aug 12];6(6):495–7. Available from: https://www.jaadcasereports.org/article/S2352-5126(20)30270-8/fulltext
17. Lymphomatoid Papulosis Treatment & Management: Medical Care, Consultations, Activity [Internet]. [cited 2020 Oct 23]. Available from: https://emedicine.medscape.com/article/1098954-treatment
18. Lymphomatoid Papulosis - Infectious Disease Advisor [Internet]. [cited 2020 Oct 23]. Available from: https://www.infectiousdiseaseadvisor.com/home/decision-support-in-medicine/dermatology/lymphomatoid-papulosis/
19. Kempf W, Pfaltz K, Vermeer MH, Cozzio A, Ortiz-Romero PL, Bagot M, et al. EORTC, ISCL, and USCLC consensus recommendations for the treatment of primary cutaneous CD30-positive lymphoproliferative disorders: lymphomatoid papulosis and primary cutaneous anaplastic large-cell lymphoma. Blood [Internet]. 2011 Oct 13 [cited 2022 Aug 12];118(15):4024–35. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3204726/
20. Liu H, Hoppe R, Kohler S, Harvell J, Reddy S, Kim Y. CD30+ cutaneous lymphoproliferative disorders: The Stanford experience in lymphomatoid papulosis and primary cutaneous anaplastic large cell lymphoma. J Am Acad Dermatol. 2004 Jan 1;49:1049–58.