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Introduction

- A 32-year-old female
- Presented with skin blemishes, without ulceration and scratching.
- These blemishes spontaneously appeared and disappeared from time to time
- There was no history of systemic B symptoms.
- The patient did not report any past medical history and no current medication in use.
- No smoking or excess alcohol drinking

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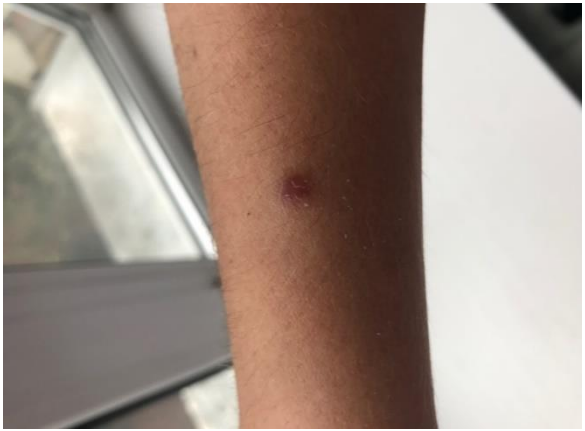
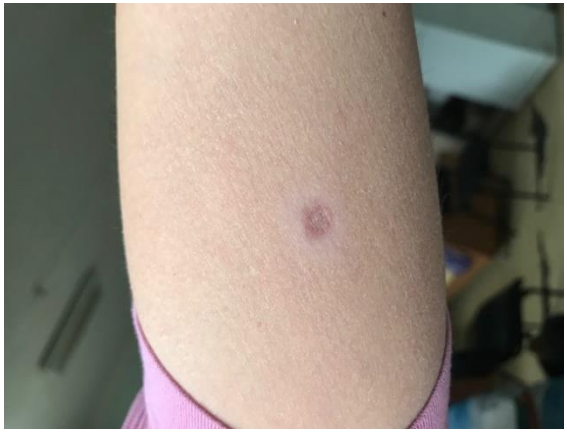
Physical examination

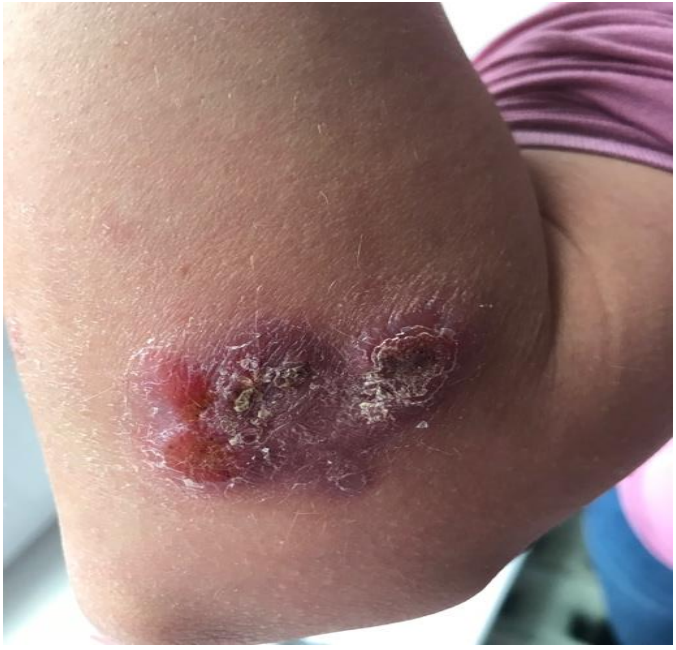




A steroid ointment was prescribed by the dermatologist

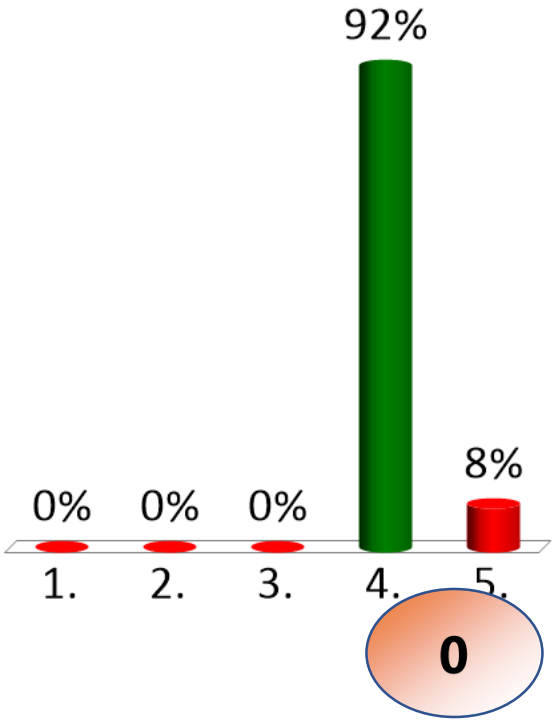
History

- Three months later she noted growing itching plaque with ulceration on the internal part of the right elbow joint.



Q1) How to proceed?

1. Watch & Wait
2. Confocal Laser Scanning Microscopy
3. Local Radiotherapy
4. Skin biopsy
5. Fine needle biopsy



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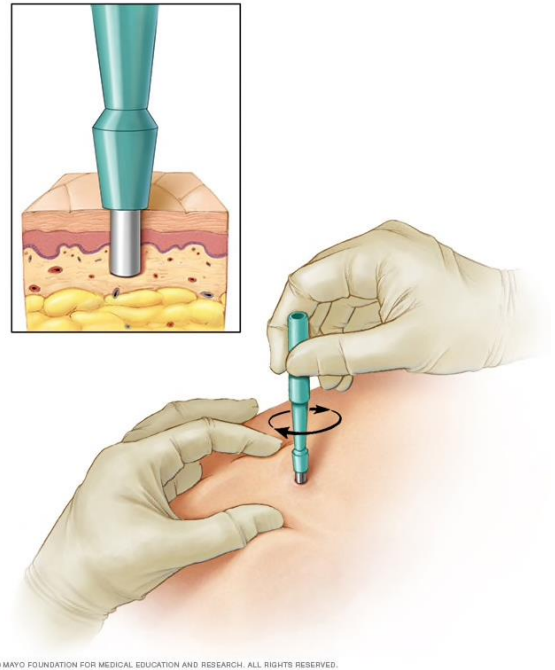
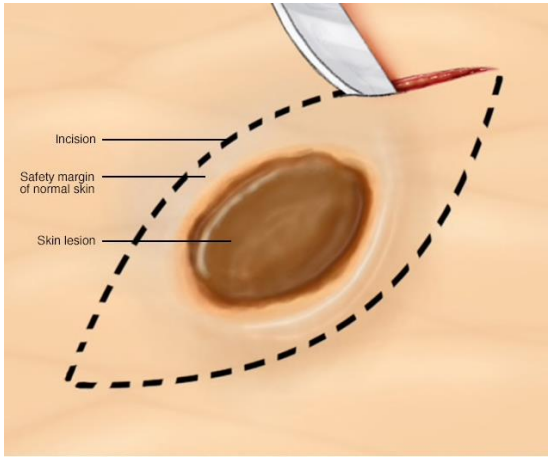
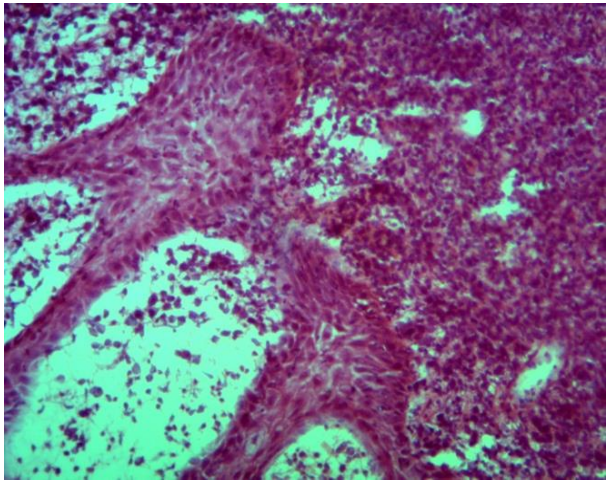
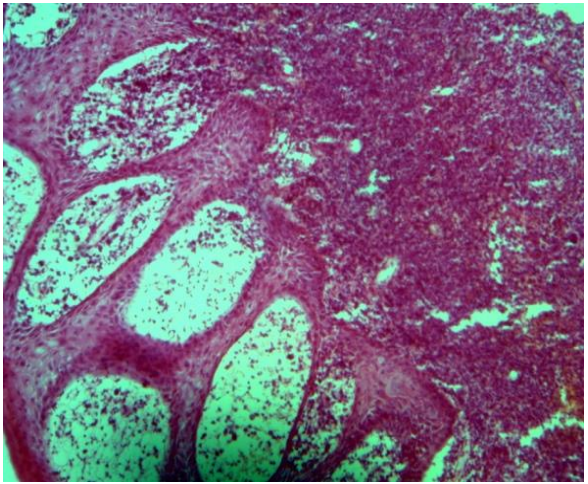
Punch and excisional biopsy





The skin biopsy was done.
In the biopsy material, there were skin fragments with diffuse infiltration by elements of lymphoid origin. Tumor showed a diffuse growth with polymorphic abnormally shaped nuclei, with light eosinophilic cytoplasm, high epidermotropism and presence of necrosis





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Immunophenotype

- CD2+, CD4+
- CD3- CD8-
- Strong CD30 positivity
- CD246 (ALK) -

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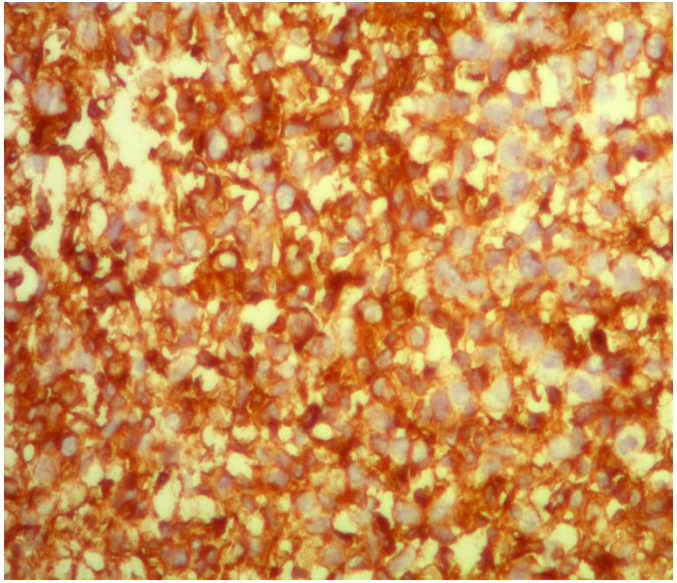
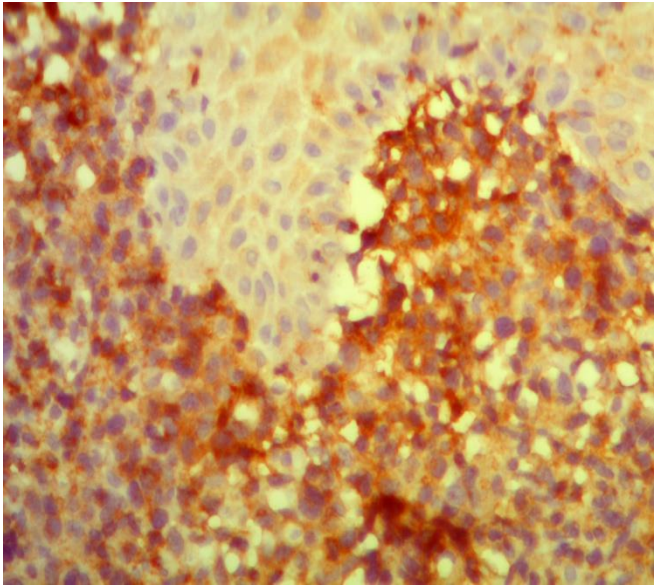
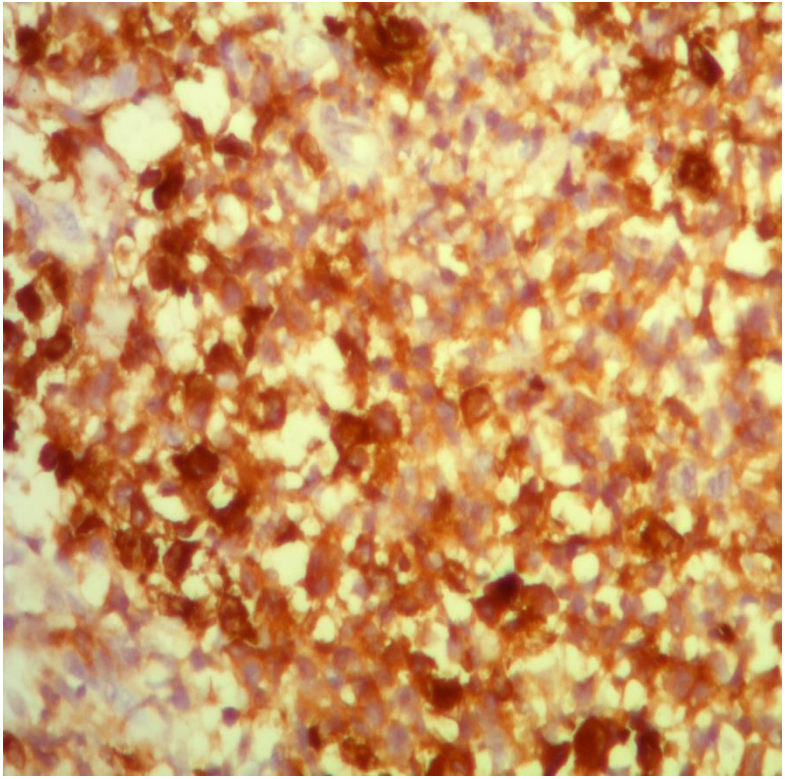
CD2



CD4

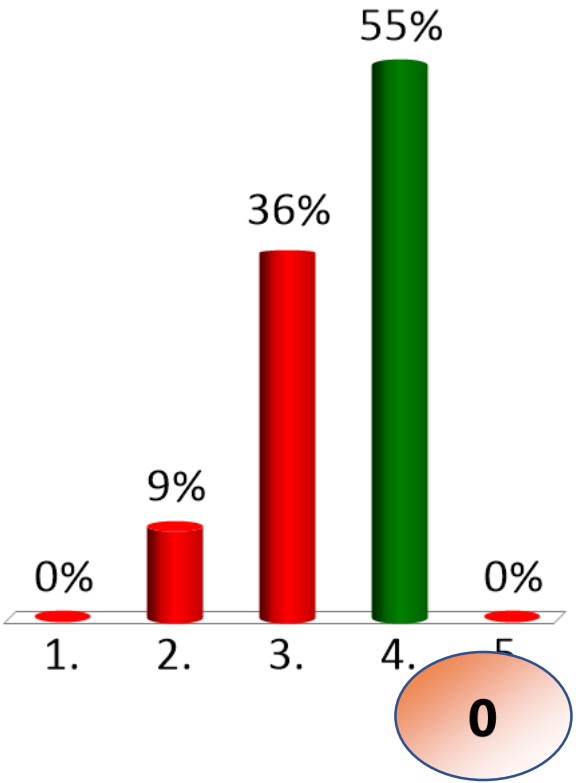




CD30

Q2: Which diagnosis?

1. Cutaneous Follicular Lymphoma
2. Sezary syndrome
3. Peripheral T Cell Lymphoma, unspecified
4. Primary cutaneous Anaplastic Large Cell Lymphoma
5. Skin carcinoma



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Staging

- Peripheral blood flow cytometry analysis showed no evidence of lymphoma or leukemia.
- Patient had normal CD4/CD8 ratio and normal expression of B-cell and T-cell antigens.
- The whole body CT-scan was negative
- No superficial lymph nodes enlargement were noted.

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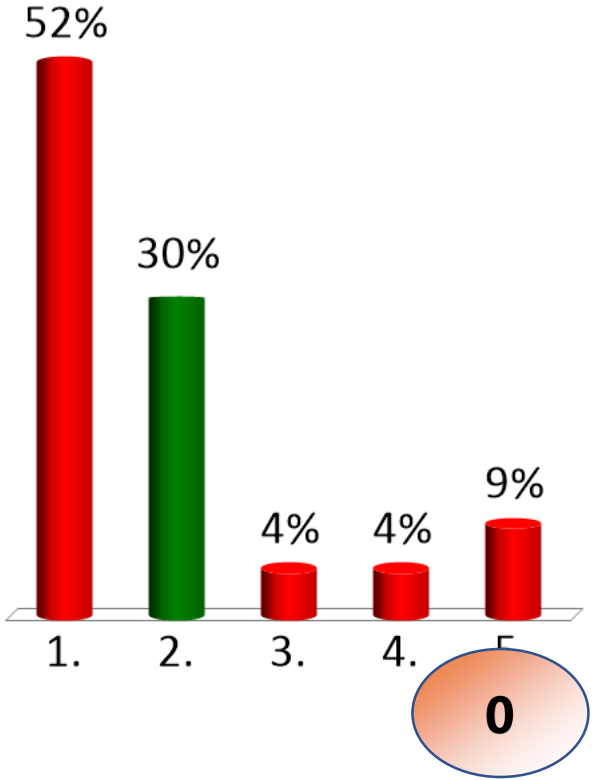
Diagnosis

Primary cutaneous Anaplastic Large cell lymphoma, ALK negative, Stage I

Q3: Which therapy?

1. CHOP or CHOEP
2. Radiation therapy
3. Steroids
4. PUVA-therapy
5. Brentuximab Vedotin



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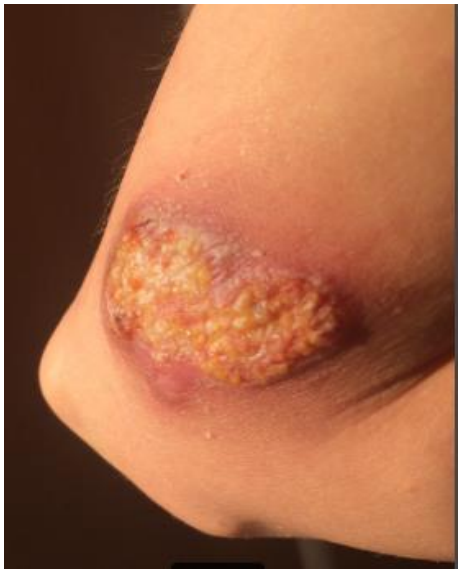
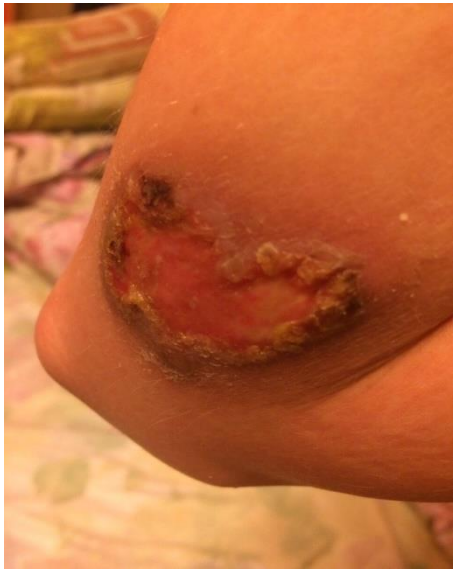
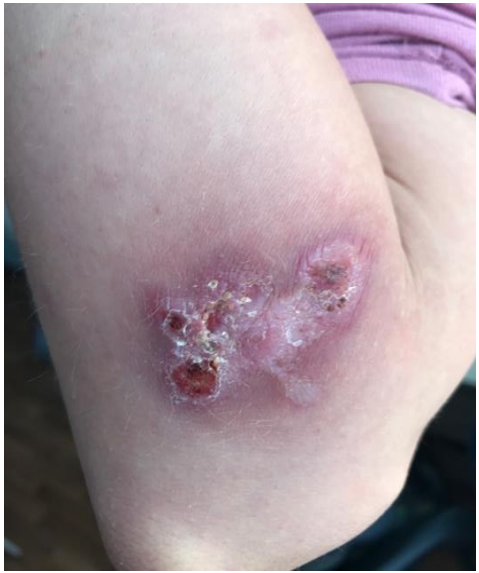
Treatment

Radiation therapy to ulcerated lesion, locally. Systemic corticosteroids and antihistamine treatment were used to resolve itching.



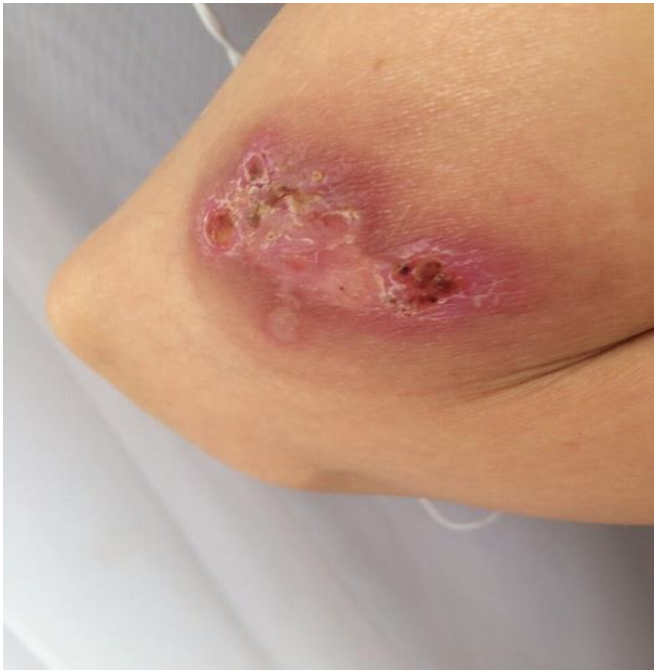




Imrpvement during RT

Treatment

A complete response was achieved after 30 Gy RT



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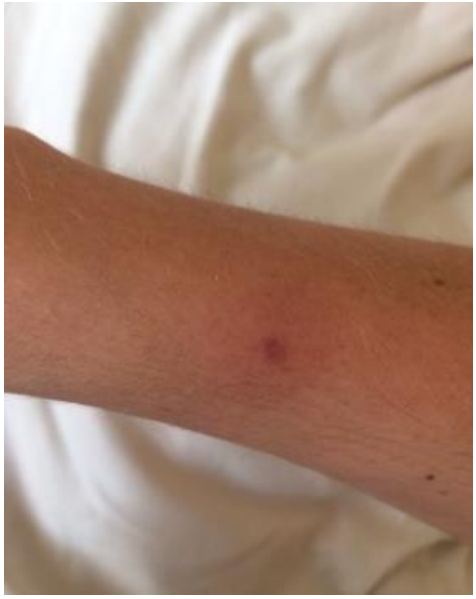
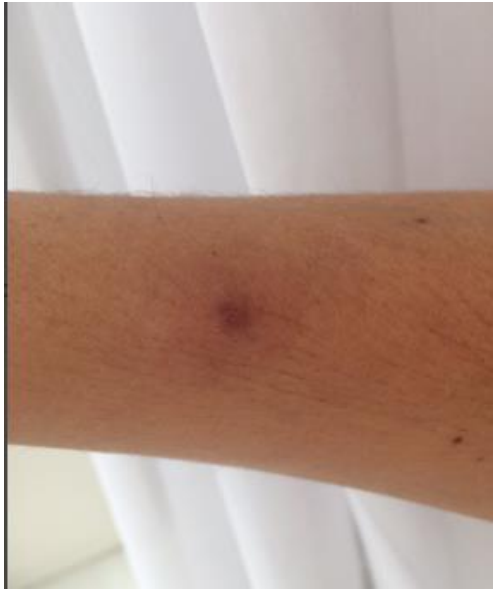
Few months later small blemishes were appearing on the skin again





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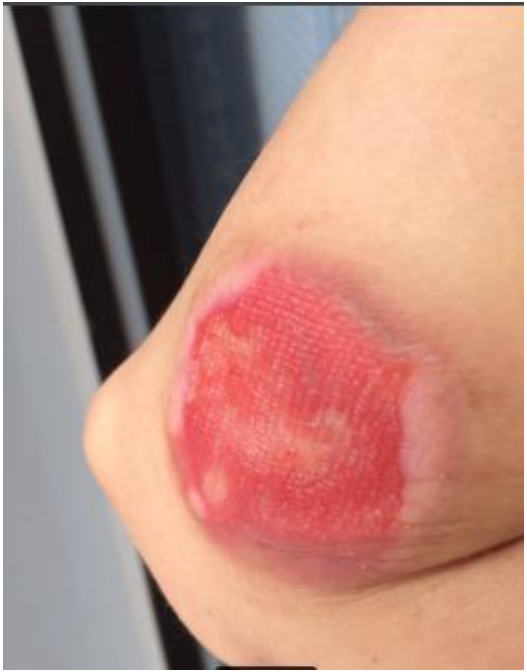
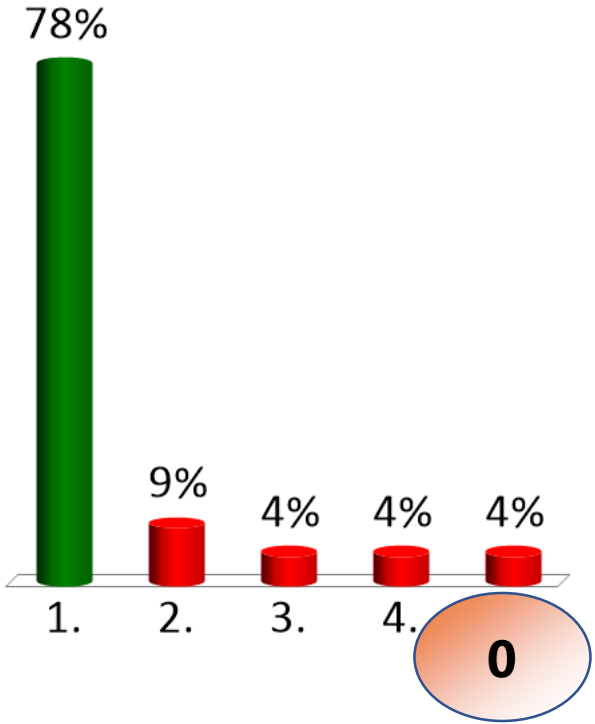
In a few weeks one of the lesion expanded ..



Q4: Which therapy?

1. CHOP or CHOEP
2. Radiation therapy
3. Low dose methotrexate
4. PUVA-therapy
5. Brentuximab Vedotin



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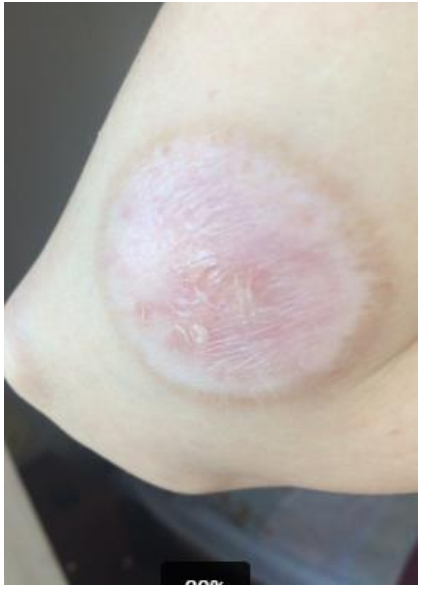
After 4 courses of CHOEP treatment was stopped



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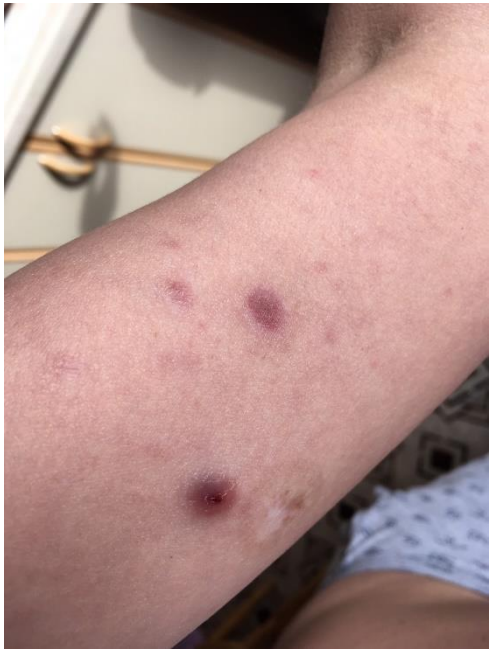
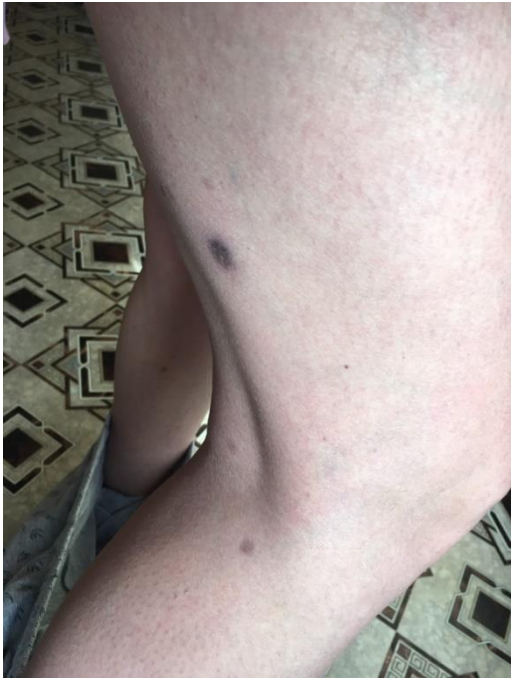
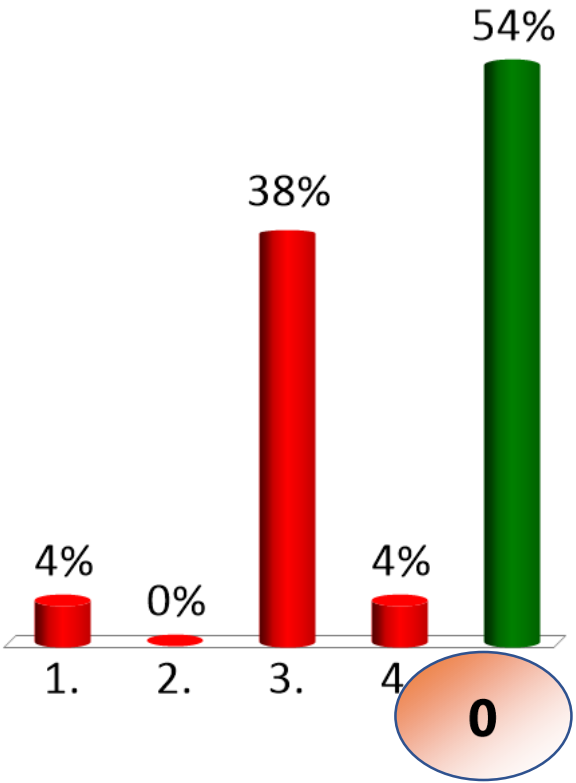
New lesions appeared few months later





Q5: Which therapy?

1. Resume CHOP or CHOEP
2. Bendamustin
3. High dose therapy + Autologous Stem Cell Transplant
4. PUVA-therapy
5. Brentuximab Vedotin



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Outcome

- Clinical assessment after the first 4 cycles showed a complete disappearance of lesions.
- She is still on BV therapy (course # 8)

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Discussion

- There are many options for treating Primary Cutaneous ALCL available, but localized treatment with either radiation therapy or surgical excision are the preferred therapies for single lesions.
- **Radiation therapy is most commonly used, and has a response rate of 100%. Radiation therapy has been shown to be be more effective and longer lasting than using multi-agent ("traditional") chemotherapy.**
- For multiple or widespread lesions in which radiation therapy or surgery aren't good options, there are several possible alternatives for treating PCALCL.

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Discussion

- For patients with relapse, systemic therapy is recommended.
- **First line systemic therapy should based on anthracicline containing regimens (CHOP or CHOEP)**
- For subsequent relapses several alternatives are now available, including the antiCD30 Brentuximab Vedotin

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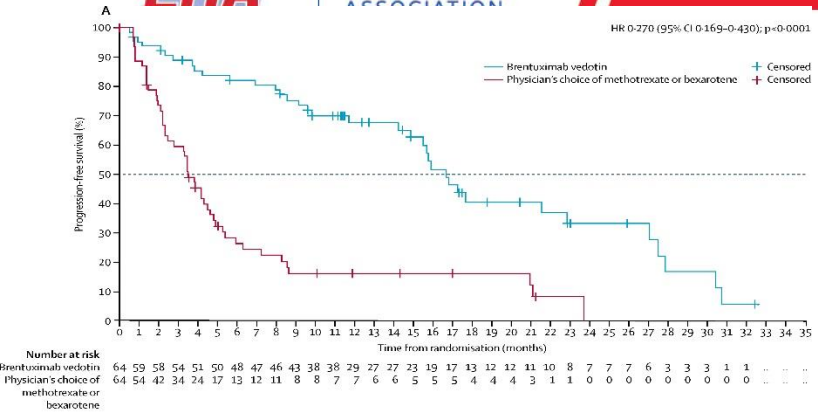

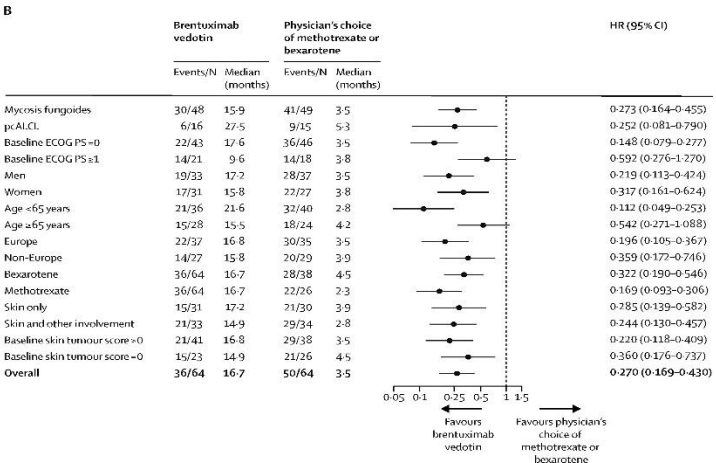

- Brentuximab vedotin or physician's choice in CD30-positive cutaneous T-cell lymphoma (ALCANZA): an international, open-label, randomised, phase 3, multicentre trial