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*Case Report*

# Kikuchi-Fujimoto disease with supraclavicular adenopathy and ribosomal P positivity in a long-standing case of discoid lupus erythematosus

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## ABSTRACT

We report a case of Kikuchi-Fujimoto disease (KFD) in a 28-year-old female with long-standing discoid lupus erythematosus. Supraclavicular lymphadenopathy in KFD is rarely reported. Recurrence of KFD and ribosomal P positivity are other unique features in this case.

**Keywords:** Discoid lupus erythematosus, Kikuchi-Fujimoto disease, Supraclavicular lymphadenopathy

## INTRODUCTION

Kikuchi-Fujimoto disease (KFD) is an uncommon idiopathic lymphadenitis that often has a self-limiting course and primarily affects young women. Cervical lymphadenopathy, with or without systemic symptoms, is the most prevalent clinical manifestation of this condition.[1] Here, we report a case of KFD associated with supraclavicular lymphadenopathy who had discoid lupus erythematosus (DLE) of 10 years duration.

## CASE REPORT

A 28-year-old female presented to the rheumatology department with fever, joint pain, fatigue, and tender swelling of her neck for two months. She was referred to the dermatology department for evaluation of patchy hair loss. She gave a history of patches of hair loss, gradually increasing in size for the past 10 years. There was no history of weight loss, night sweats, photosensitivity, oral ulcers, or diffuse hair loss.

General examination revealed multiple tender, firm, mobile, discrete 1-2 cm sized supraclavicular lymph nodes on the right side and posterior auricular and cervical lymph nodes on both sides. She also had multiple depressed patches of alopecia on the vertex, frontal, and parietal areas with size varying from 3 × 3 to 4 × 2 cm with adherent scales [Figure 1a and b]. There were no other skin lesions. There was no hepatosplenomegaly. There was no joint swelling or restriction of movements.

A dermoscopy of the scalp showed follicular plugging, white structureless areas with a periphery showing hyperpigmentation, scaling, and reduced follicular ostia [Figure 2a]. Biopsy from the

scalp showed epidermal thinning with follicular plugging and minimal periappendageal inflammatory infiltrates, basal cell vacuolar degeneration of the epidermis and follicular epithelium, and pigment incontinence in the dermis suggestive of DLE. Direct immunofluorescence showed granular staining of the basement membrane zone with Immunoglobulin G, Immunoglobulin M, Immunoglobulin A, and C3. An ultrasonogram of lymph nodes showed right supraclavicular and bilateral cervical lymphadenopathy with preserved fatty hilum [Figure 2b]. A biopsy of the lymph node showed necrotizing lymphadenitis with histiocytic collection with nuclear debris [Figure 2c]. There were no well-formed granulomas or malignant cells. The hemogram, peripheral smear, thyroid function test, and chest X-ray were normal. Viral markers, Interferon-Gamma Release Assay, and tissue Cartridge-Based Nucleic Acid Amplification test were negative. Antinuclear antibody (ANA) test, anti-ds Deoxyribonucleic acid antibody, and direct coombs test were negative. C3 and C4 (complement) levels were normal.

A final diagnosis of KFD with long-standing DLE was made. The patient was treated with hydroxychloroquine 200 mg twice daily and antipyretics. There was complete arrest in the progression of scalp lesions with improvement of systemic symptoms. She was under regular follow-up with no clinical evidence of systemic lupus erythematosus (SLE).

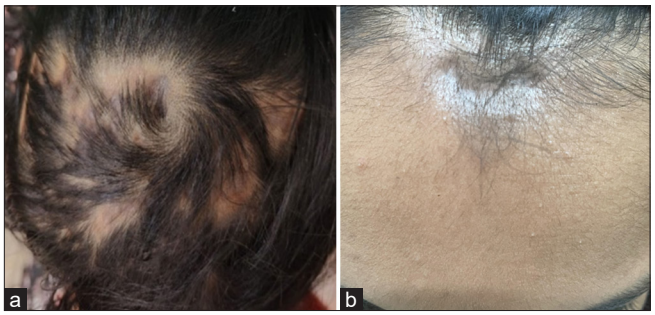


**Figure 1 (a):** Multiple depressed hyperpigmented alopecic plaques on the vertex and parietal with adherent scales; (b): Depressed alopecic plaque on the frontal area.

After 6 months, she presented with a recurrence of swelling of supraclavicular lymph nodes and was reevaluated. ANA immunoblot showed strong positivity to Ribosomal P (Rib P) antigen. She was then given systemic steroids in tapering doses along with hydroxychloroquine, which resulted in the resolution of the swelling, and is under follow-up.

## DISCUSSION

Kikuchi-Fujimoto's necrotizing lymphadenitis (cervical subacute necrotizing lymphadenitis or histiocytic necrotizing lymphadenitis) was first described in 1972 by Kikuchi and Fujimoto independently.[1]

Our patient had scarring alopecia due to DLE for 10 years without any other features of SLE. There has been a single case report of KFD after 10 years of DLE, reported by Silver *et al*.[1] According to a long-term study conducted from January 1990 to December 2010, 31% of KFD patients eventually progressed to develop definite SLE, while 8% had only lupus-like syndrome.[2]

Tender lymphadenopathy, which typically affects the cervical lymph nodes, is one of the clinical symptoms at presentation of KFD. In more severely affected patients, this condition is also linked to fever, weight loss, and night sweats.[2] Even though cervical lymphadenitis is common in KFD, supraclavicular lymphadenopathy is extremely rare, with only one case reported by Sarfraz *et al*.[3] Apart from SLE, other diseases that might cause KFD include lymphoma, drug eruptions, viral lymphadenitis, and tuberculous lymphadenitis.[2] Hence, a thorough workup to exclude these conditions is mandatory in all cases of KFD.

Although our case went into remission initially, she came with a relapse of symptoms after 6 months and was reevaluated. Recurrence in KFD is reported only in 3–4% of cases.[4] There was a recurrence of swelling of lymph nodes, which were reassessed sonologically. She was also found to have a Rib-P-positive ANA profile. Patients who

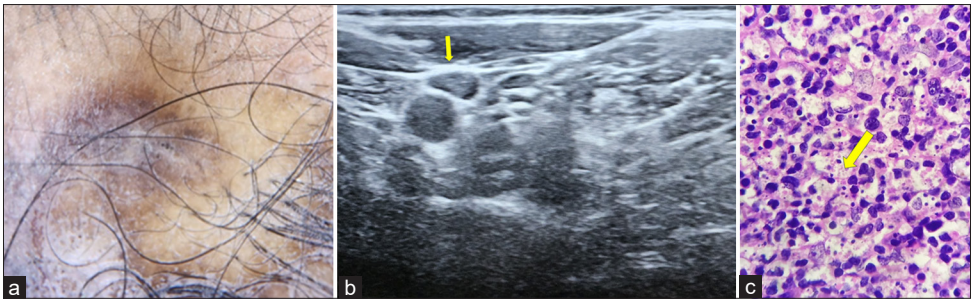


**Figure 2 (a):** Dermoscopy showing follicular plugging, white structureless areas with periphery showing hyperpigmentation, scaling, and reduced follicular ostia (DermLite DL3N Dermoscope); (b): ultrasonogram lymph nodes showing enlarged lymph nodes with preserved fatty hilum (yellow arrow); and (c): necrotizing lymphadenitis histiocytic collection with nuclear debris (yellow arrow) (hematoxylin and eosin, 40x).

have anti-Rib-P antibodies tend to have a reduced age at disease onset, involvement of multiple organs, and an overall severe course of the disease with central nervous system involvement.[5] Although she does not fulfill the European Alliance of Associations for Rheumatology (EULAR)/ Systemic Lupus International Collaborating Clinics (SLICC) criteria for SLE, the recent development of anti-Rib-P antibody suggests possible progression to SLE.

Many patients receive supportive care with non-steroidal anti-inflammatory drugs to reduce related symptoms such as fever, pain from lymphadenitis, or arthralgia. Corticosteroids are utilized in several studies to treat severe forms of this disease (not responding to non-steroidal anti-inflammatory drugs, with protracted course). Intravenous immunoglobulin and hydroxychloroquine are also successfully used to treat incurable diseases.[5]

## CONCLUSION

We report KFD in a case of long-standing DLE, which had unusual features like supraclavicular adenopathy, which is largely under-reported. On further follow, she also developed a recurrence of KFD associated with anti-Rib-P antibody positivity.

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## REFERENCES

1. Goodfield M, Dutz J, McCourt C. Lupus erythematosus In: Griffiths EM, Barker J, Bleiker T, Chalmers R, Creamer D, eds. Rook's textbook of dermatology (9th ed). UK: WILEY Blackwell; 2016.p. 51.31.
2. James JF, Liles JE, Thakore MW. Lupus profundus and discoid lupus following a diagnosis of Kikuchi-Fujimoto disease. JAAD Case Rep 2021;33:45-7.
3. Sarfraz S, Rafique H, Ali H, Hassan SZ. Case report: Kikuchi-Fujimoto disease: A case of supraclavicular lymphadenopathy. F1000Res 2019;8:1652.
4. Hyun M, So IT, Kim HA, Jung H, Ryu SY. Recurrent Kikuchi's disease treated by hydroxychloroquine. Infect Chemother 2016;48:127-31.
5. Carmona-Fernandes D, Santos MJ, Canhão H, Fonseca JE. Anti-ribosomal P protein IgG autoantibodies in patients with systemic lupus erythematosus: Diagnostic performance and clinical profile. BMC Med 2013;11:98.

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