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Kikuchi Fujimoto Disease associated with Cerebral palsy- Rare histiocytic necrotizing lymphadenitis

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Conflicts of Interest: Nil

Abstract

Aim and objectives: To study the case of Kikuchi Fujimoto Disease associated with Cerebral palsy and arouse the awareness about the disease.

Materials And Methods: A 27-year-old female patient presented with bilateral multiple swelling over the neck, loss of weight, loss of appetite and low-grade fever since one and half months. On local examination,

patient had bilateral level I II III IV and V enlarged lymph nodes which were non tender, non-inflamed, discrete, non-matted, firm in consistency. ESR and CRP levels were raised. Patient's USG neck revealed multiple level lymph nodes which were showing fatty hilum and few showing necrosis with internal echos. FNAC was suggestive of atypical lymphoid hyperplasia. Patient underwent lymph node biopsy and a sample sent for HPE.

Results: HPE Reported the lymph node shows paracortical zone of necrosis. Karyorrhectic and karyolytic debris are seen. Neutrophils are very scanty showing Kikuchi necrotizing lymphadenitis.

Discussion

- Lymphadenopathy is one of the most common clinical problems encountered in paediatrics.[2]
- This is a rare occurrence of KFD disease in cerebral palsy patient.

Conclusion: This highlights the importance of keeping kikuchi Fujimoto disease (histiocytic necrotizing lymphadenitis) in the differential diagnosis of fever with cervical lymphadenopathy in young female.

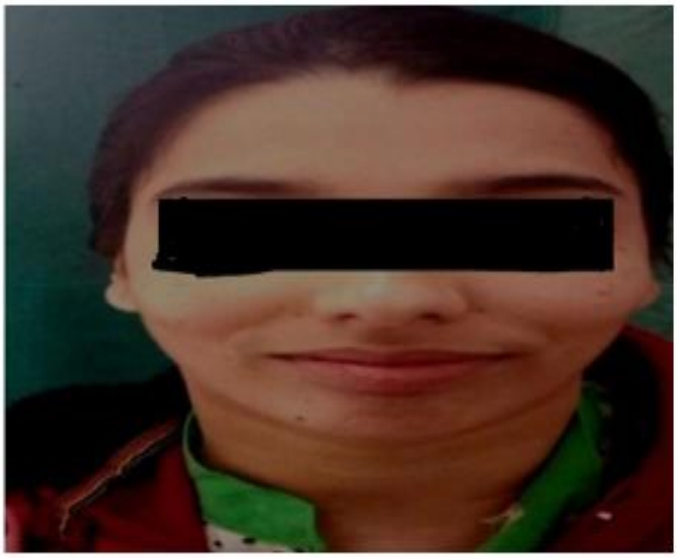
Keywords: Kikuchi Fujimoto Disease (KFD), Cervical Lymphadenopathy, Cerebral palsy, young age, SLE

Introduction

Lymphadenopathy is one of the most common clinical problems encountered in paediatrics.[2] The precise incidence of lymphadenopathy is not known, but estimates of palpable adenopathy in childhood vary from 38% to 45%.[1] There are approximately 300 lymph nodes in the neck and they have considerable capacity to undergo growth and change.[3] With their high concentration of lymphocytes and antigen-presenting cells, lymph nodes are ideal for receiving antigens that gain access through the skin or gastrointestinal tract. They are barely perceptible in neonates, but a progressive increase in antigen exposure will lead to an increase in lymph node size until later childhood. Lymph node atrophy will start during adolescence and continues through later life. This heterogeneous group of benign

lymphoproliferative disorders is characterized by prolonged unexplained cervical adenopathy which can give rise to concerns regarding as 'pseudo lymphomata'. They include Rosai–Dorfman disease, Castleman disease, Kawasaki syndrome and Kikuchi-Fujimoto disease.[4]

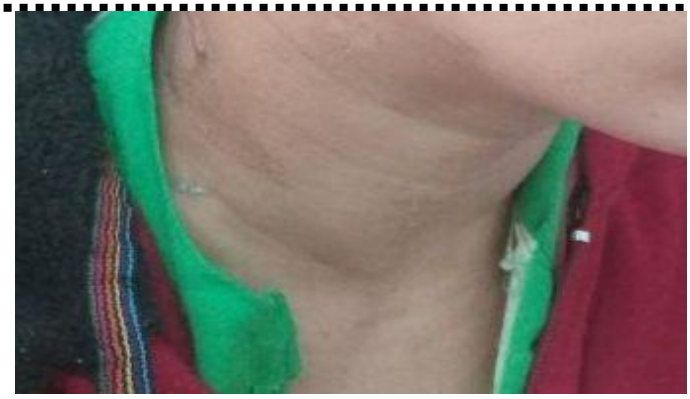
Kikuchi-Fujimoto disease was first described by Kikuchi from Japan in 1972.[5,6] It is an idiopathic and rare condition which is also known as **histiocytic necrotizing lymphadenitis**. It has high prevalence in Asian and Japanese people. Fever chills, tender cervical lymphadenopathy and weight loss are common. Women and young adults are more commonly affected. The disease is self-limiting but biopsy is often performed to rule out malignancy. Histology shows a characteristic necrotizing lymphadenitis.



Case Report

- 27 years old female k/c/o cerebral palsy
- Patient had bilateral swelling over the neck
- h/o loss of weight
- h/o loss of appetite
- Low grade fever since 1 n ½ months.
- O/E: Bilateral level Ia Ib II III IV V lymph nodes were palpable

Investigations



- Hemoglobin 12.1gm/dl , TLC -8300/cumm
- Platelet count 271*1000
- CXR PA view normal
- USG Abdomen WNL
- ANA and dsDNA negative
- Interferon gamma assay negative
- ESR and CRP are mildly elevated.
- Mantoux test was negative.
- Serology testing was negative.

FNAC

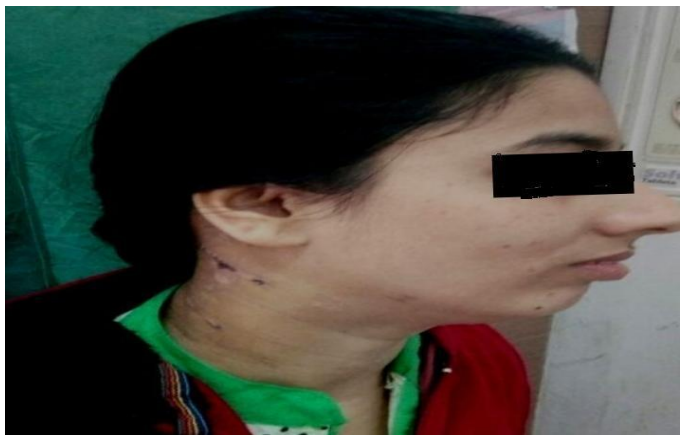
It was showing atypical lymphoid hyperplasia and few histiocytes.

USG neck

- B/L levels I II III IV and V lymph nodes show fatty hilum and few of them showing necrosis with internal echoes suggestive of necrosis within.

Diagnosis

Patient underwent a right sided level V excisional lymph node biopsy.

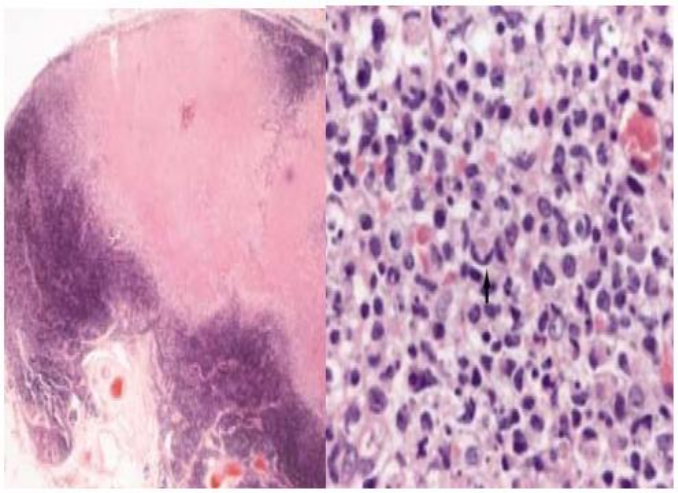


Histopathology

Macroscopy: 3*3 cm sized lymph node was well circumscribed, ovoid shaped. smooth surfaced with regular margins and it had a rubber like consistency.

Microscopy: the lymph node showed a paracortical zone of necrosis. Karyorrhectic and karyolitic debris were seen. Neutrophils were very scanty. Histiocytes with eccentric crescent shaped nucleus had monocytic appearance.

Careful histopathological examination helps us distinguish from SLE is the almost total absence of plasma cells in the nodal tissue. From lymphoma includes incomplete architectural effacement with patent sinuses, presence of reactive histiocytes, low mitotic rates and absence of Reed Sternberg cell.



Treatment

Symptomatic treatment -NSAIDS were given.

Discussion

Young female patient with a known case of Cerebral palsy presented with bilateral swelling over the neck, loss of weight, loss of appetite and low-grade fever with since one and half months. On local examination, patient had bilateral level I II III IV and V enlarged lymph nodes which were non tender, non-inflamed, discrete, on matted, firm in consistency. ESR and CRP levels were raised. Patient's USG neck revealed multiple level lymph nodes which were showing fatty hilum and few showing necrosis with internal echoes.

Serologic testing for antibodies against these infections has consistently shown negative results, and no viral particles have been found[7].

Serologic testing of antinuclear antibodies (ANA) has been found consistently negative for KFD[7] .

Careful histopathological examination helps us distinguish from SLE is the almost total absence of plasma cells in the nodal tissue. From lymphoma includes incomplete architectural effacement with patent sinuses, presence of reactive histiocytes, low mitotic rates and absence of Reed Sternberg cell.

HPE Reported the lymph node shows paracortical zone of necrosis. Karyorrhectic and karyolytic debris are seen. Neutrophils are very scanty showing kikuchi necrotizing lymphadenitis.

This is a rare occurrence of KFD disease in cerebral palsy patient.

Conclusion

This case demonstrates the importance of excisional biopsy in recognizing KFD.

This disorder must be considered among the differential diagnosis when a young patient presents with fever and cervical lymphadenopathy.

Recognition of this condition is crucial, because it can be mistaken for malignant lymphoma or SLE. Awareness of this disorder not only by clinicians but also by pathologists might help prevent wrong diagnosis of KFD and harmful and unnecessary evaluations and treatments of patients.

References

1. Locke R, MacGregor F, Kubba H. The validation of an algorithm for the management of paediatric cervical lymphadenopathy. Int J PediatrOtorhinolaryngol 2016; 81:5-9.

2. Grossman M, Shiramizu B. Evaluation of lymphadenopathy in children. Curr Opin Pediatr 1994; **6**(1): 68–76.
3. Trotter HA. The surgical anatomy of the lymphatics of the head and neck. Ann Otol Rhinol Laryngol 1930; **39**: 384–97.
4. Brown JR, Skarin AT. Clinical mimics of lymphoma. Oncologist 2004; **9**: 406–16.
5. Phelan E, Lang E, Gormley P, Lang J. Kikuchi-Fujimoto disease: a report of 3 cases. Ear Nose Throat J 2007; **86**(7).
6. Payne JH, Evans M, Gerrard MP. Kikuchi- Fujimoto disease: A rare but important cause of lymphadenopathy. Acta Paediatr 2003; **92**(2): 261–4.
7. Tariq H, Gaduputi V, Rafiq A, Shenoy R: The enigmatic Kikuchi-Fujimoto disease: a case report and review. Case Rep Hematol. 2014, 2014:648136. 10.1155/2014/648136