KIKUCHI’S DISEASE: A CLINICOPATHOLOGICAL ANALYSIS OF 22 CASES

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ABSTRACT

Objective: To determine the clinicopathological pattern of Kikuchi’s disease.

Material and Methods: This case series study was conducted at the department of Ear, Nose, Throat, Head and Neck Surgery, Postgraduate Medical Institute, Lady Reading Hospital, Peshawar from Jan. 2002 to Dec. 2008. This study includes 22 cases of Kikuchi’s disease. Patients of any age and either sex presenting to E.N.T Department and diagnosed as Kikuchi’s disease were included in this study. These patients were treated with oral prednisolone, antibiotics and analgesics and they were followed at 3 monthly intervals for at least 12 months. The data of these patients were collected on a preformed proforma. The statistical analysis was performed using the statistical program for social sciences (SPSS version 11).

Results: Our study included 22 cases of Kikuchi’s disease constituting 13 females and 09 males, with female: male ratio of 1.4:1. The age of the patients ranged from 11-47 years. Most of the patients presented in their 2nd and 3rd decade, followed by patients in their 4th and 5th decade. The clinical features of these patients were mainly lymphadenopathy and fever. The diagnosis of Kikuchi’s disease was made on the basis of fine needle aspiration cytology in 18 cases (81.81%) and 4 cases (18.18%) were diagnosed on lymph node histopathology. These patients were treated with oral prednisolone and antibiotics.

Conclusion: While dealing with a young patient with fever and cervical lymphadenopathy, the clinicopathological profile of Kikuchi’s disease must be kept in mind both by clinicians and pathologists to prevent misdiagnosis.

Key Words: Kikuchi’s disease, Histiocytic necrotizing lymphadenitis, Cervical lymphadenopathy.

INTRODUCTION

Kikuchi-Fujimoto disease (KFD) or Histiocytic necrotizing lymphadenitis is a rare, benign and self-limiting disease of unknown etiology, which is usually characterized by cervical lymphadenopathy, fever and leukopenia. This disease was first reported by Kikuchi and Fujimoto et al in 1972 in Japan. Kikuchi’s disease is known to have a worldwide distribution. The description of this disease outside Asia was made by Pilieri and colleagues in 1982 who reported 23 cases from the former West Germany and individual cases from Iran, Italy, South Korea, and Spain. The etiology of Kikuchi’s disease is not known. Various etiological agents like human herpes virus (HHV6 and HHV8), herpes simplex virus, adenovirus, parvovirus B19, cytomegalovirus, varicella zoster, dengue virus, bacteria such as Mycobacterium azulgai, yersinia and protozoa have been linked to the disease. The onset of Kikuchi’s disease is acute or subacute, evolving over a period of 2 to 3 weeks. Cervical lymphadenopathy is present in 56% to 98% of cases, more commonly consisting of tender lymph nodes involving the posterior cervical triangle and generally unilateral. Generalized lymphadenopathy has been reported in 1% to 22% of cases. Involvement of mediastinal, peritoneal, and retroperitoneal regions is uncommon. In addition to lymphadenopathy, 30% to 50% of patients with Kikuchi’s disease might have fever, usually of low-grade, nausea, vomiting, sore throat and night sweats. Systemic symptoms are found more frequently when extranodal involvement is present. The usefulness of fine-needle aspiration cytology (FNAC) to establish a cytologic diagnosis of Kikuchi’s disease has been accepted. However excisional lymph node biopsy should be mandatory if clear-cut clinical and cytologic Kikuchi’s disease findings are absent. This disease does not have a characteristic appearance on ultrasonographic or computed tomographic (CT) examination because the findings of CT and magnetic resonance imaging of Kikuchi’s disease can mimic that of metastatic and tuberculous lymph nodes. The characteristic histopathologic findings of Kikuchi’s...
disease include irregular paracortical areas of coagulative necrosis with abundant karyorrhectic debris, which can distort the nodal architecture, and large numbers of different types of histiocytes at the margin of the necrotic areas. The differential diagnosis of Kikuchi’s disease mainly includes reactive lesions such as lymphadenitis associated with systemic lupus erythematosus (SLE), herpes simplex and other microorganisms, non Hodgkin lymphoma, plasmacytoid T-cell leukemia, Kawasaki disease, nodal colonization by acute myeloid leukemia and even metastatic adenocarcinoma. The aim of this study is to identify the burden of this clinical condition in our set-up.

**MATERIAL AND METHODS**

This descriptive case series study was conducted at the Department of Ear, Nose, Throat, Head and Neck Surgery, Postgraduate Medical Institute, Lady Reading Hospital, Peshawar from January 2002 to December 2008. Patients of all ages and either sex presenting to ENT department and diagnosed as Kikuchi’s disease were included in this study. The patients with cervical lymphadenopathy due to other causes were excluded from the study. The diagnosis of this disease was obtained by fine needle aspiration cytology in the majority of cases and some of the cases were diagnosed by subsequent excisional biopsy of lymph nodes. All these patients were evaluated in terms of detailed history, thorough examination and relevant investigations. These patients were treated with oral prednisolone in the dose of 1mg/kg body weight for a maximum of 3 months duration in tapering dose and combined with antibiotics and analgesics for 2-4 weeks and they were followed at 3 monthly intervals for at least 12 months. The data of these patients were collected on a preformed proforma. The statistical analysis was performed using the statistical program for social sciences (SPSS version 11). The frequencies and percentages were presented for qualitative variables and Mean ± SD were presented for quantitative variables.

**RESULTS**

Our study included 22 cases of Kikuchi’s disease, 13 female and 09 male, with female: male ratio of 1.4:1. The age of the patients ranged from 11-47 years. Most of the patients presented in the 2nd and 3rd decade followed by the 4th and 5th decade. The clinical features of these patients were lymphadenopathy, fever and anemia (Table 1). The cervical lymph nodes were multiple in majority of cases with the right side of neck involved. The size of the lymph nodes ranged from 2-7.2 cm with a mean of 4.40 cm. The specific investigations carried out in this study were ultrasonography, fine needle aspiration cytology and histopathology of the lymph node.

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<tr>
<th>S. No.</th>
<th>Symptoms</th>
<th>No. of cases &amp; percentage</th>
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<tbody>
<tr>
<td>1</td>
<td>Neck swelling</td>
<td>22(100%)</td>
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<tr>
<td>2</td>
<td>Fever</td>
<td>17(77.27%)</td>
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<tr>
<td>3</td>
<td>Sore throat</td>
<td>09(40.90%)</td>
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<tr>
<td>4</td>
<td>Body aches</td>
<td>06(27.27%)</td>
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<thead>
<tr>
<th>Signs</th>
<th>No. of cases &amp; percentage</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Cervical lymphadenopathy</td>
</tr>
<tr>
<td>2</td>
<td>Axillary lymphadenopathy</td>
</tr>
<tr>
<td>3</td>
<td>Anemia</td>
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<tr>
<td>4</td>
<td>Weight loss</td>
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Table 1: Clinical Features of the Patients (n=22)

Fig. 1: Picture of a patient with Kikuchi’s disease having scar of lymph node biopsy in the right posterior triangle of neck.

Fig. 2: Micrographic picture of Kikuchi’s disease showing Karyorrhectic foci with large numbers of histiocytes (Hematoxylin & Eosin stain).
The diagnosis of Kikuchi’s disease was made on the basis of FNAC in 18 cases (81.81%) and 4 cases (18.18%) were diagnosed by histopathology of lymph node (Fig. 2). These patients were treated with oral steroids and antibiotics.

**DISCUSSION**

Kikuchi’s disease is a self-limiting benign systemic lymphadenitis of unknown cause especially involving the cervical nodes. This disease was reported originally by Kikuchi and Fujimoto in Japan and is now considered to be a global disease existing in Pakistan also. The female to male ratio in our study was 1.4: 1., simulating other studies showing female predominance. The reason for this is not clear but female hormones may play some role as reported by Seo JH, and others. This disease is more common in younger people as described by most of the authors. In our study, Kikuchi’s was found predominantly in the 2nd and 3rd decade of life which is in accordance with other studies. The clinical profile of this disease constitutes lymphadenopathy, fever and body aches. In our study lymphadenopathy was seen in all patients. The cervical lymph nodes (100%) were involved which is in accordance with study of Bosch X and others. Similarly fever was the second most common (77.27%) presentation in our study. This is also reported by Babu NC and colleagues. The rare presentations of these patients include headache, body aches, vomiting and weight loss. In our study some of the rarer complaints were body aches (27.27%) and weight loss (22.72%). Similar complaints were also reported by many authors and in our study some of the rarer complaints were body aches (27.27%) and weight loss (22.72%). The clinical features of Kikuchi’s disease simulate other conditions like systemic lupus erythematosus (SLE), lymphoma and immunological disorders so proper diagnosis is important because this is a self limiting condition.

In our study Kikuchi’s disease was diagnosed by FNAC (81.81%) and by histopathology (18.18%) which is also in accordance with other studies. This is a benign condition with good response to steroids as reported by many authors. Majority of the patients in our study responded to oral steroids, antibiotics and analgesics. They were regularly followed and had no recurrence. Literature on this disease is reviewed extensively and major papers are case reports except few one and according to our knowledge it is first case series study in Pakistan. The clinicopathological profile is presented to help in early diagnosis and treatment of patients of Kikuchi’s disease.

**CONCLUSION**

Although Kikuchi’s disease is an extremely uncommon possibility when dealing with a young patient with fever and cervical lymphadenopathy, the clinicopathological profile of this disorder must be kept in mind both by clinicians and pathologists to prevent misdiagnosis and inappropriate treatment.

**REFERENCES**


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