

CRPS is a term to describe conditions predominantly characterized by spontaneous or stimulus-induced pain that is inconsistent with the provocative event [3]. CRPS diagnosis entirely depends on observable signs and reported symptoms, which have been put together into various diagnostic criteria sets for adults [4]. The diagnosis of CRPS in children may be delayed as long as four months because of the low incidence and considerably different clinical presentation compared with adults [5]. The clinical manifestations of CRPS may imitate rheumatologic diseases in children.

CRPS has been suggested to be a multifactorial condition that is related to an unusual host response to certain tissue damage. The disease often includes a wide diversity of autonomic and motor disturbances like hyperalgesia, allodynia, and sensory loss [4,5]. The patient can exhibit particularly painful, red, warm, and swollen extremities, mimicking trauma. Other probable accompanying features are changes in sweating, reduced hair and nail growth, allodynia and hyperalgesia, and also muscle weakness [2]. However, apparent trauma or initiating factor is absent in most of the pediatric patients. Exclusion of other possible causes is necessary and investigations are needed to exclude infections of skin, connective tissues, muscles, bone and joints. Further investigations may be needed if inflammatory diseases, fractures, neoplasms and deep venous thrombosis are suspected [6]. In our patient, the possible conditions were excluded by further clinical and laboratory evaluation.

The optimal management approach should consist a multidisciplinary treatment of noninvasive interventions including physiotherapy, occupational therapy, analgesics and psychotherapy [5,6]. Sensory rehabilitation is sometimes added in order to gradually improve the allodynia [6]. The standard medications include drugs non-steroidal anti-inflammatory drugs, antidepressants,

anticonvulsants, topical analgesic patches. In contrast to adults, the response to treatment, particularly exercise therapy with behavioral management will achieve almost 97% remission in children [3], as in the reported child.

We report a pediatric CRPS Type I patient, treated successfully by conservative methods, in order to attract attention to this rare benign condition in children.

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Extra-nodal Kikuchi Disease and Kikuchi Encephalitis

Kikuchi disease is a rare but benign, self-limiting disease that typically presents with prolonged fever and cervical lymphadenopathy. We report neurological manifestations in an adolescent girl with recurrent Kikuchi disease.

Keywords: *Lymphadenopathy, Meningoencephalitis, Necrotizing lymphadenitis, Recurrence.*

Kikuchi-Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis has a recurrence rate of around 3-7% in reports from most centers but can range as high as 20% in predisposed East-Asian populations [1]. Its pathogenesis still remains controversial [2]. Neurological manifestations of Kikuchi disease are extremely rare and isolated, and include headache, peripheral neuropathy, aseptic meningitis to encephalitis [3,4].

A 14-year-old girl presented to our children's emergency with fever for 10 days. She was reviewed by a pediatrician who prescribed antibiotics for bacterial tonsillitis. In view of persistent fever and neck swelling, she consulted our emergency department. On physical examination, she had right-sided cervical lymphadenopathy (5 x 5 cm). She had a history of KFD when she was 9 years old and presented similarly. She required excision of the affected lymph nodes and the biopsy showed necrotizing histiocytic lymphadenitis. There were concerns of aseptic meningitis during that episode as she had worsening headaches with neck stiffness and giddiness. Neuro-imaging showed prominent meningeal enhancement but cerebrospinal analysis was normal. She was treated with a course of indomethacin and steroids during that episode.

During her current admission, her blood investigations revealed mildly elevated inflammatory markers with raised C-reactive protein (CRP) of 44 mg/L and erythrocyte sedimentation rate (ESR) of 59 mm/h. She was treated initially for infective cervical lymphadenitis with intravenous amoxicillin and clavalanate. Alternative differentials were considered as her fever persistent despite adequate antibiotic coverage. During the fourth day of hospitalization, she was noted to be acutely confused. In view of fluctuating consciousness, she was transferred to intensive care unit (ICU) for further management and subsequently intubated in view of encephalopathy. Antibiotic therapy was escalated to intravenous Ciprofloxacin with addition of Acyclovir. Neuro-imaging did not reveal any abnormalities. Spinal tap was done with opening pressure at 52 cm H₂O. Cerebrospinal fluid (CSF) was noted to be turbid with fluid analysis showing significant pleocytosis and raised protein (CSF WBC 60/ μ L, total protein 0.9 g/L). In view of concerns of possible Kikuchi-related encephalitis, pulse methylprednisolone for 5 days with subsequent tapering doses of steroids were administered. During her stay in ICU, she was noted to be increasingly agitated with sleep-cycle disturbances. An electroencephalogram (EEG) did not reveal any epileptiform activity or electrographic seizure. CSF and blood investigations were not suggestive of N-Methyl-D-Aspartic acid (NMDA) encephalitis. An extended autoimmune screen was negative for any concerns of underlying autoimmune encephalitis. Infective screening was negative for Bartonella, Epstein-Barr virus, Mycoplasma, Tuberculosis, HIV, Leptospirosis, Melioidosis, Toxoplasmosis and Rickettsia. She underwent lymph node excision biopsy, which showed necrotizing histiocytic lymphadenitis. She was diagnosed to be having a recurrence of KFD with encephalitis. On recovery, she had mild in-coordination with aphasia. There were other neuropsychiatric symptoms of attention-deficit, increased lability and sleep disturbances during her recovery. Her cognition was appropriate, and she was able to return to school within 6 weeks post discharge.

KFD has a very good prognosis with remission occurring spontaneously in most cases. Some cases require a course of steroids or non-steroidal anti-inflammatory drugs (NSAIDs).

KFD is an uncommon condition in the pediatric population and extra-nodal manifestations are uncommon [5]. Our patient had recurrent KFD, and on both occasions, there was varying degrees of neurological manifestations. Extra-nodal Kikuchi disease should be considered as a differential in a child with acute encephalopathy if there were preceding symptoms of lymphadenopathy with prolonged fever.

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