Arch Pediatr Infect Dis.2013;1(1):36-39. DOI: 10.5812/pedinfect.5351

Archives of Pediatric Infectious Diseases
www.pedinfect.com
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Tuberculous Pachymeningitis in a Young Child With Spinal Involvement: A Case Report

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ARTICLE INFO

Article type:
Case Report

Article history:
Received: 29 Apr 2012
Revised: 12 May 2012
Accepted: 27 May 2012

Keywords:
Tuberculosis
Child
Meningitis

ABSTRACT

Pachymeningitis is a rare chronic disorder of the duramater caused by various infectious, autoimmune or malignant diseases. We report a child with chronic tuberculous pachymeningitis, who presented with clinical manifestations of meningitis with a compatible cerebrospinal fluid analysis. Despite signs of progressive neurological involvement, extensive work-up done to rule out known causes of dural inflammation was negative. The patient was started empirically on anti-tuberculous therapy, to which he responded after 2 weeks. He was discharged on anti-TB medications. He remains well on follow-up. We recommend a trial of anti-tuberculous treatment in children presenting with signs of pachymeningitis in whom the cause of chronic meningeal inflammation cannot be identified.

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Implication for health policy/practice/research/medical education:
This article describes a case of pachymeningitis, this condition has largely been reported in adults, with very few reports in children.

Please cite this paper as:
Karimi A, Rafiei Tabatabaei S, Shiva F, Sayfikar M, Jafari M, Shirvani F, et al. Tuberculous Pachymeningitis in a Young Child With Spinal Involvement: A Case Report. *Arch Pediatr Infect Dis*.2013;1(1): 36-9. DOI: 10.5812/pedinfect.5351

1. Introduction

Pachymeningitis is a rare disorder characterized by inflammation and thickening of dura mater which may result in neurological deficits. This condition has largely been reported in adults, with very few reports in children (1-6).

The present report describes a 4 year old boy with pachymeningitis, his course of illness, and his response to treatment.

2. Case Description

A 4 year old boy was hospitalized with fever, abdominal pain and vomiting that began 4 weeks ago and had been treated with metoclopramide, ranitidine, omeprazole and cefotaxime, with no improvement. During this period he had lost 5Kg.

On admission to the hospital he looked ill, with an axillary temperature of 38.4°C, pulse rate 110/min and a blood pressure of 115/75 mm Hg. He revealed signs of meningeal irritation, viz. a stiff neck and positive Kernig's and Brudzinski's signs. Fundoscopy revealed bilateral papilledema, rest of the neurological examination was normal. Abdominal examination was normal except for mild tenderness in the peri-umblical region. Heart and lungs were normal.

Results of Lab test were as follows: Hemoglobin: 11.8 mg/

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DOI: 10.5812/pedinfect.5351
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dL, white blood count (WBC): 14900/mm^3, neutrophils (PMN): 67%, lymphocytes (L): 30%, monocytes: 2%, eosinophils: 1%, platelets: 558000/mm^3, erythrocyte sedimentation rate: 80 mm/hr, C-reactive protein: 30 mg/dL, blood glucose: 156 mg/dL. Serum albumin, electrolytes and tests for renal, liver and thyroid function were normal. Chest x-ray reported only multiple calcified lymph nodes in the right axilla. Tuberculin test (PPD), was negative. Cerebrospinal fluid (CSF), analysis was as follows: WBC: 250/mm^3, (PMN: 40%; L:60%) RBC: 0/mm^3, Protein: 42 mg/dL, glucose: 20 mg/dL; cultures were negative after 72 hours.

He was started on intravenous Ceftriaxone and Vancomycin and corticosteroids with a diagnosis of chronic meningitis. Normal results were found for: anti-nuclear antibody, Perinuclear Anti-Neutrophil Cytoplasmic Antibodies, Cytoplasmic Anti-Neutrophil Cytoplasmic Antibodies, Complements (C3, C4, CH50), Anti double stranded DNA. Serology for Human Immunodeficiency virus (HIV), *Human T-lymphotropic virus* (HTLV), leishmaniasis, toxoplasmosis, brucellosis, and cytology and polymerase chain reaction (PCR) for tuberculosis (TB) in the CSF were negative. CSF Indian ink test for cryptococcosis was negative. CSF PCR for *Cytomegalovirus* (CMV), *Herpes simplex*, (HSV), and *Epstein-Barr virus* (EBV) were negative. Results of Bone marrow aspiration were normal. Brain magnetic resonance imaging (MRI) was requested which revealed meningeal enhancement in the frontal lobe consistent with pachymeningitis (*Figure* 1).

Four days after admission, he developed hemiparesis on the left side. Spinal cord MRI was requested which revealed a mass lesion consistent with a neurogenic tumor in nerve roots at T10-T11 level on the right side.

The patient was transferred to another hospital for neurological surgery where he had a generalized seizure followed by loss of consciousness. The spinal mass was operated and removed. Magnetic resonance Spectrometry of the brain reported extensive signal abnormality in both frontal lobes in the cortical and sub cortical regions and in the left periventricular white matter. Biopsy of the mass revealed a chronic granulomatous lesion within fibrotic stroma with bundles of nerve fibers and ganglion cells, but no malignancy. The picture was compatible with tuberculosis (*Figure* 2). On repeated questioning no history of contact with tuberculosis was obtained. Simple and nested PCR of paraffin embedded block for *Mycobacterium tuberculosis* from extradural mass T10-T11 was done which was negative.

Empiric anti-tuberculosis treatment was started, he slowly regained consciousness and his general condition improved. Four weeks after starting anti tuberculosis (TB) treatment, results of CSF analysis were as follows: Glucose 45 mg/dL; protein 110 mg/dL; RBC 10/mm^3 and WBC 3/mm^3 and cultures were negative. Five days later his fever subsided. He was discharged on anti-tuberculosis therapy with regular follow up.

3. Discussion

Pachymeningitis was first described by Joffroy and Charcot in 1869; most cases in those days were attributed to either tuberculosis or syphilis (1, 2). Nafzigger later described a patient with what he named as Idiopathic Hypertrophic Pachymeningitis, (IHP) as no definite cause could be determined for the inflammation of duramater (2). In addition to syphilis, and tuberculosis, chronic meningitis is seen in other bacterial, viral, Spirochetal, parasitic and fungal infections. It has also been reported in association with malignancy, rheumatic and autoimmune disorders like rheumatoid arthritis, Behcet's Disease, Wegener granulomatosis, temporal arteritis, Fabry disease, central nervous system vasculitis and also in sarcoidosis. Furthermore, various drugs viz. NSAIDs, intrathecal agents, and intravenous immunoglobulin have been implicated in causing chronic meningeal inflammation (7-12). About 5% of patients with sarcoidosis may have neurological involvement, which may be the presenting sign in some of the cases (8). In many cases no definite etiology can be determined; IHP, therefore is a diagnosis of exclusion which is made on the results

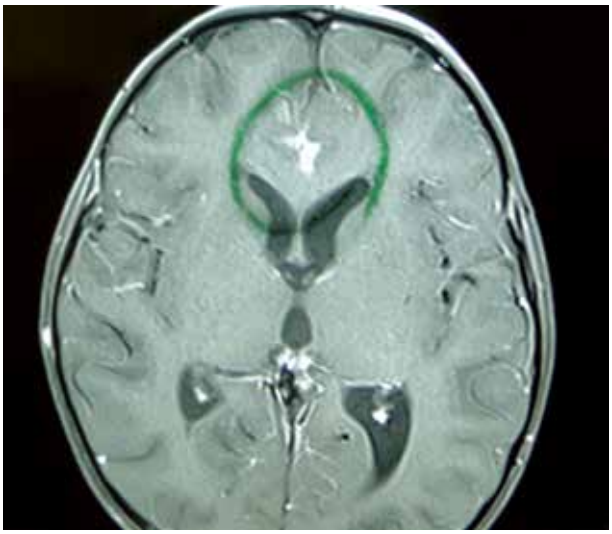

Figure 1. Brain MRI Revealing Leptomeningeal Enhancement in Frontal Lobe, Consistent With Pachymeningitis

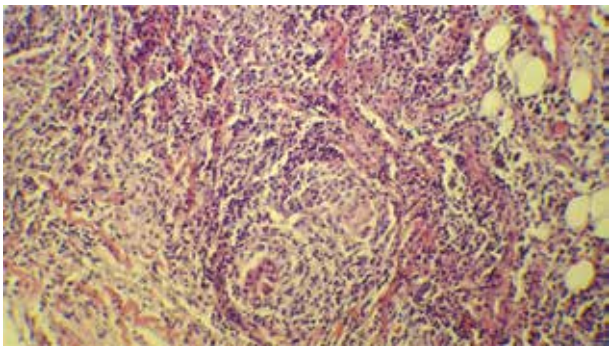

Figure 2. Biopsy of the Spinal Mass Shows Granulomatous Lesion

of a meningeal biopsy that excludes known causes (3). Aburahma *et al*. have reported the development of pachymeningitis in a 3.5 year old child following head trauma; a diagnosis of IHP was made by meningeal biopsy and by ruling out other known causes (5). Another case of IHP in a 10-year old child presenting with polyneuropathies was reported by van Troom *et al*.; their patient responded well to systemic corticosteroids and oral methotrexate (6).

Clinical manifestations in pachymeningitis are caused by compression of adjacent structures by the thickened and inflamed duramater (2). These may include chronic headaches, ataxia, facial pain, cranial nerve involvement and neuro-ophthalmic complications such as papilledema and various neurological deficits. Ophthalmic involvement with 6th nerve palsy retrobulbar optic neuropathy and scleritis, have been reported in adults with pachymeningitis (9, 10).

Results of all diagnostic tests were negative in our patient; initially he was started on antibiotics and corticosteroids with presumptive diagnoses of chronic infectious meningitis or IHP, however his general condition worsened and he developed signs of increasing neurological involvement.

After the results of the spinal biopsy which revealed a chronic granulomatous disease compatible with tuberculosis and because of worsening signs and repeated hypoglycorrhachia, empiric antituberculous therapy with rifampin, isoniazid, pyrazinamide, and ethambutol was begun which resulted in marked improvement of all his symptoms. *Mycobacterium tuberculous* infection presenting as pachymeningitis is uncommon, but this phenomenon has been reported previously in adults (13-16).

It is often difficult to make a definite diagnosis of tuberculous meningitis as isolating *Mycobacterium tuberculosis* from the spinal fluid by acid fast staining, culture or even PCR is difficult (15). An increase in the adenosine deaminase level in the CSF has been used to clinch the diagnosis of TB, especially to differentiate it from meningeal sarcoidosis which may have a similar presentation (15). This laboratory test however, was not performed and we relied on the biopsy reports and the response to the anti-Tb treatment to establish the diagnosis.

Tuberculous meningitis is the most severe form of extrapulmonary tuberculosis; because of difficulty and delay in diagnosis; , also because delayed treatment results in high morbidity and mortality, many experts recommend prompt empiric therapy (16). On the basis of our experience with this case we recommend that anti-tuberculous treatment should be started in children presenting with pachymeningitis due to unidentified causes before making a diagnosis of idiopathic hypertrophic pachymeningitis. This is especially important in countries where childhood tuberculosis is prevalent.

Acknowledgments

None declared.

Authors' Contribution

F. Shiva: Data review, Literature search and writing and revising the paper.

A Karimi and S Rafiei Tabatabaei: Involved in acquisition and interpretation of the data and revision of manuscript.

R Shiari: Helped in data gathering and review of manuscript.

F shirvani, M Seyfikar and M Jafari contributed in data collections.

Financial Disclosure

None declared.

Funding/Support

None declared.

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