Case Report

A Case of JRA with Tuberculosis Hypertrophic Cranial Pachymeningitis

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Abstract

Hypertrophic cranial pachymeningitis is a rare inflammatory disease with diffused involvement of the dura and also is a rare disorder of diverse etiology. Based on anatomic site cases of hypertrophic pachymeningitis can be subdivided into spinal, intracranial and craniospinal pachymeningitis. In this study, we present a 21 years old woman with Tuberculosis pachymeningitis who had juvenile rheumatoid arthritis since infancy too. She developed headache and seizure 4 and 2 months ago respectively. Her headache became severe and she experienced 2 attacks of generalized tonic-clonic seizures. Dural biopsy showed neither rheumatoid nodules nor vasculitic process. Considering clinical features such as morning stiffness, arthralgia and joint tenderness and absence of clinical and laboratory evidences of infectious causes, MRI findings were most consistent with rheumatoid pachymeningitis. Based on clinical features, MRI, and laboratory test results, rheumatoid pachymeningitis was suspected for the patient and IV methylprednisolone was administered. Finally, this report emphasizes the importance of considering TB as one of the erythematous meningitis that is rarely encountered in RA [1]. The imaging features of hypertrophic cranial pachymeningitis include dural thickening, dural mass, sinus thrombosis, venous congestion with white matter changes. In some cases, even after extensive investigation, no specific cause is found and the process is called idiopathic intracranial pachymeningitis. Herein we describe a case of Tuberculous hypertrophic cranial pachymeningitis who also suffered from juvenile rheumatoid arthritis [2].

Case report

Here we present a 21 years old woman with Tuberculosis pachymeningitis who had juvenile rheumatoid arthritis since infancy too. She developed headache and seizure 4 and 2 months ago respectively. Her headache became severe and she experienced 2 attacks of generalized tonic-clonic seizures. The latter attack was secondarily generalized and preceded by Jacksonian focal motor seizure of right upper and lower extremities added to these symptoms they were fever and intermittent focal motor seizures of right leg that developed few weeks before admission. N/E revealed hyperreflexia of right lower extremity. Laboratory findings showed leukocytosis and elevated ESR and positive CRP. RA titer and ANA were negative. The CSF study including bacteriologic examination was normal. T1 weighted MRI demonstrated hypertrophy of cranial dura extending from falx cerebi to cerebral convexities which were enhanced markedly by GD-DTPA. The dura adjacent to the cavernous sinus was also thickened and enhanced (Figure 1,2,3,4). Dural biopsy showed neither rheumatoid nodules nor vasculitic process. Considering clinical features such as morning stiffness, arthralgia and joint tenderness and absence of clinical and laboratory evidence of infectious causes, MRI findings were most consistent with rheumatoid pachymeningitis. Based on clinical features, MRI, and laboratory test results, rheumatoid pachymeningitis was suspected for the patient and IV methylprednisolone was administered. After transient initial improvement the patient developed abdominal pain, N/V, and decreased loss of consciousness. Abdominal sonography revealed multiple enlarged para aortic lymph nodes that after surgical biopsy showed granulomatous reaction in favor of TB. Subsequent anti-TB treatment resulted in clinical improvement and the patient was discharged after regain of consciousness and resolution of neurological impairments.

Discussion

HCP is a rare disease with chronic inflammation of dural matter which may cause neurological symptoms secondary to compression of adjacent neural structures. The most common clinical symptoms in cranial pachymeningitis are increased Intracranial pressure and consequently, chronic headache, cranial nerve palsies (i.e. ophthalmoplegia, hypoacusia and dysphagia) [3].

It can also cause cerebellar ataxia, optic neuropathy and even blindness [1]. Radiculopathy or myopathy are among common manifestations of spinal pachymeningitis [4]. HCP occurs with symptoms that can mimic other diseases including subdural...
hemorrhage and dural carcinomatosis. CT shows the affected dura as a high-density lesion that enhanced after contrast administration. MRI demonstrates an isointense lesion with remarkable contrast enhancement on the T1-weighted image and a hypointense lesion with a hyperintense rim on the T2-weighted image indicating a dense fibrous tissue and inflammatory infiltrates, but even its appearance in MRI can be very similar to diseases like meningioma en plaque, lymphoma, TB and sarcoidosis, so when the systemic work up was not diagnostic, dural biopsy via craniotomy is suggested [5]. HCP is divided into two primary (Idiopathic) and secondary types. The most common secondary causes of which are infectious causes including TB and syphilis which are far more common in developing countries and specially in immunocompromised patients. Other causes of secondary HCP can be called inflammatory diseases (i.e Sarcoidosis, Rheumatoid Arthritis, Wegner Granulomatosis and Polyarteritis nodosa), traumatic and toxic causes. In some cases, even after extensive studies no specific cause can be found for HCP, that in these cases it is called idiopathic HCP. Rheumatoid pachymeningitis is a rare occasion in RA. Both fibrinoid deposits and rheumatoid nodules can cause pachymeningitis.

During the onset of neurological disease, symptoms of active synovitis may or may not be present. CSF analysis may show high protein concentration in TB pachymeningitis. Although mycobacterium tuberculosis DNA has been reported positive in various studies, but its negative result cannot be the basis for ruling out the disease. A definitive diagnosis of TB pachymeningitis is by PCR and histopathologic examination. But despite of all of this, many cases of TB pachymeningitis are detected only through response to anti TB drug therapy [1]. Just as our patient in who was identified in this way. This topic emphasizes the importance of considering TB as one of the most common causes of pachymeningitis especially in endemic area despite the existence of negative PCR and PPD tests.

Conclusion

This report emphasizes the importance of considering TB as one of the most common causes of pachymeningitis especially in the endemic area despite the existence of negative PCR and PPD tests.
References


Figure 3: Axial view without Gad.

Figure 4: Axial view with Gad.