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Rare presentation of tuberculous hypertrophic pachymeningitis diagnosed by a biopsy of abdominal lymphadenopathy

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Abstract

A 59-year-old man with medical history of diabetes mellitus and hypertension presented with a persistent fever of unknown origin and developed a headache. Laboratory tests, including polymerase chain reaction assays for *Mycobacterium tuberculosis*, showed no specific abnormal findings in blood or cerebrospinal fluid. Contrast-enhanced computed tomography revealed abdominal paraaortic lymphadenopathy. Abdominal lymph node biopsy showed caseous necrosis and suggested tuberculous lymphadenopathy. Intensive examinations revealed positive T-SPOT.TB test and multiple dural nodular hypertrophic lesions in brain magnetic resonance imaging. After antitubercular treatment, all clinical manifestations and dural nodular lesions improved. Finally, we diagnosed the patient with tuberculous hypertrophic pachymeningitis. To our knowledge, this is the first report of tuberculous hypertrophic pachymeningitis concomitant with abdominal tuberculous lymphadenopathy and no other dissemination. Systematic investigation of tuberculosis is important for pachymeningitis.

Keywords

Tuberculous hypertrophic pachymeningitis, abdominal paraaortic lymphadenopathy, tuberculous lymphadenopathy

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Introduction

Hypertrophic pachymeningitis is a rare chronic fibrotic inflammatory disease characterized by localized or diffuse thickening of the dura mater, leptomeninges, and tentorium. Clinical symptoms include headache, vomiting, cranial nerve palsy, ataxia, raised intracranial pressure, and focal neurological deficits.[1] Among its main associated diseases are trauma, infection, autoimmune diseases, connective tissue diseases, sarcoidosis, and malignancy diseases, sarcoidosis, and malignancy.[2]

Tuberculous infection outside the lungs is not easy to diagnose, because it usually presents with an insidious clinical presentation.[3]

Lymphadenopathy is a common extrapulmonary manifestation of tuberculosis; among tuberculous lymphadenopathies, cervical tuberculous lymphadenopathy is common, while abdominal paraaortic lymph node tuberculosis is quite rare.[4] Despite the frequent occurrence of these manifestations, they pose a diagnostic problem since they mimic other pathological processes and have inconsistent physical and laboratory findings.[5] Diagnosis of such cases is difficult and often requires biopsy.

We describe a rare case of intracranial hypertrophic pachymeningitis with tuberculous lymphadenopathy around the abdominal aorta.

Case report

A 59-year-old man with a past medical history of diabetes mellitus and hypertension presented with fever of unknown origin once or twice a year since 2014. He had no contact history with an individual with active tuberculosis, nor previous history of having tuberculosis. Laboratory tests

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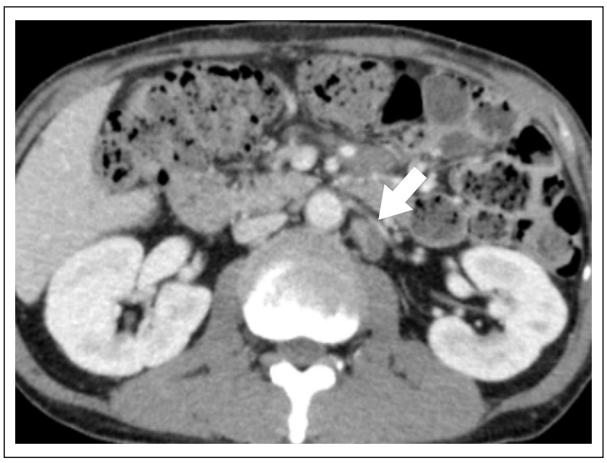

Figure 1. Contrast-enhanced computed tomography image of the abdomen showing paraaortic lymphadenopathy with homogeneous enhancement (arrow).

revealed leukocytosis with neutrophil dominance and increased serum C-reactive protein levels, otherwise cerebrospinal fluid (CSF) test revealed normal. The cause of the fever could not be identified. In 2018, since the fever had sustained, prednisolone (10 mg/day) was started under the presumption of a diagnosis of a collagen disease. In May 2019, he also presented with frontal to retro-orbital headaches. Polymerase chain reaction (PCR) analysis for *Mycobacterium tuberculosis* was negative in blood and CSF samples. Contrast-enhanced computed tomography (CT) of the abdomen performed to screen malignant neoplasms revealed lymph node swelling around the abdominal aorta (Figure 1). Because of possible malignant neoplasm, lymph node biopsy was performed. Clinicopathological analysis of an abdominal lymph node biopsy revealed caseous necrosis, and then tuberculous lymphadenopathy was suggested. Tissue culture and acid-fast staining had not been performed.

In October 2019, he visited our hospital for further examination due to headache, fever, and weight loss. Neurological examination revealed no deficits, except for meningeal signs. Laboratory tests revealed leukocytosis with neutrophil dominance and increased serum C-reactive protein levels. Serological test findings were negative for rheumatoid factor, antinuclear antibodies, IgG subclass 4, antineutrophil cytoplasmic antibodies (myeloperoxidase or proteinase-3), HIV antibody test, and tumor markers. Lumbar puncture revealed an opening pressure of 300 mm CSF. CSF analysis revealed the cell count of 3/mm^3 with lymphocyte dominance, CSF proteins 49 mg/dL, sugar 107 mg/dL, and CSF to plasma glucose ratio 0.44. No fungi, bacteria, or tuberculosis was detected in cell culture. Although PCR findings were still negative for *Mycobacterium tuberculosis* in blood and CSF samples, the T-SPOT.TB test (Oxford Immunotec, Ltd., Abingdon, UK) which is an interferon-γ release assay yielded positive results. Brain magnetic resonance imaging (MRI) demonstrated meningeal nodular hypertrophic lesions in the left dura mater, right cerebellar tentorium, and cerebral falx. Contrast-enhanced brain MRI showed homogeneous enhancement of these lesions (Figure 2(a)–(c)). Pathological specimens of biopsied lymph nodes were reviewed, and caseous necrosis with Langerhans-type giant cells, without atypical cells, was confirmed.

Antitubercular combined therapy was initiated, which comprised isoniazid (5 mg/kg/day), rifampicin (10 mg/kg/day), ethambutol (15 mg/kg/day), pyrazinamide (30 mg/kg/day), and levofloxacin (500 mg/day) for 4 months, followed by a further 10 months' administration of rifampicin (10 mg/kg/day) and isoniazid (5 mg/kg/day). Prednisolone (20 mg/day) was used for all periods of antitubercular therapy. His headache and fever improved rapidly and all clinical symptoms disappeared without sequelae for 1 year after treatment. Brain MRI after treatment demonstrated decreasing of nodular lesions in the left dura mater, right cerebellar tentorium, and cerebral falx (Figure 2(d)–(f)). The control abdominal CT had no significant change. Finally, we diagnosed tuberculous hypertrophic pachymeningitis disseminated from abdominal paraaortic tuberculous lymphadenopathy.

Discussion

This was a rare case of tuberculous hypertrophic pachymeningitis disseminated from abdominal paraaortic tuberculous lymphadenopathy. The imaging findings of tuberculous hypertrophic pachymeningitis are reported as follows: they appear isointense to brain parenchyma on T1-weighted MR images and isointense to hypointense on T2-weighted images.[6] The imaging findings of leptomeningeal involvement are not specific for tuberculosis which include carcinomatosis, lymphoma, meningioma, sarcoidosis, and syphilis.[1] We diagnosed based on pathological findings and favorable response to antitubercular treatment. Abdominal paraaortic tuberculous lymphadenopathy is a rare comorbid condition of extrapulmonary tuberculosis.[4] The most common manifestations of central nervous system (CNS) tuberculosis include tuberculous meningitis, tuberculoma, tuberculous abscess, and spinal cord tuberculosis.[7] In contrast to the frequent leptomeningeal involvement due to tubercular inflammation, pachymeningeal tuberculosis is notably uncommon and difficult to diagnosis, often requiring biopsy.[2] To our knowledge, this is the first reported case with tuberculous hypertrophic pachymeningitis and abdominal paraaortic lymphadenopathy.

Two studies have reported on tuberculous hypertrophic pachymeningitis diagnosed by cervical lymph node biopsy.[8,9] Miyamoto et al.[8] reported a case of tuberculous cervical lymphadenopathy presenting with hypertrophic pachymeningitis, he suggested *Mycobacterium tuberculosis* dissemination via the lymphatic system because lymphatic vessels are abundantly distributed in the dural sheath.[10] Suárez-Calvet et al.[9] reported a case of a patient with persistent headache for 3 years who was diagnosed as tuberculous hypertrophic pachymeningitis by caseous necrosis in the cervical

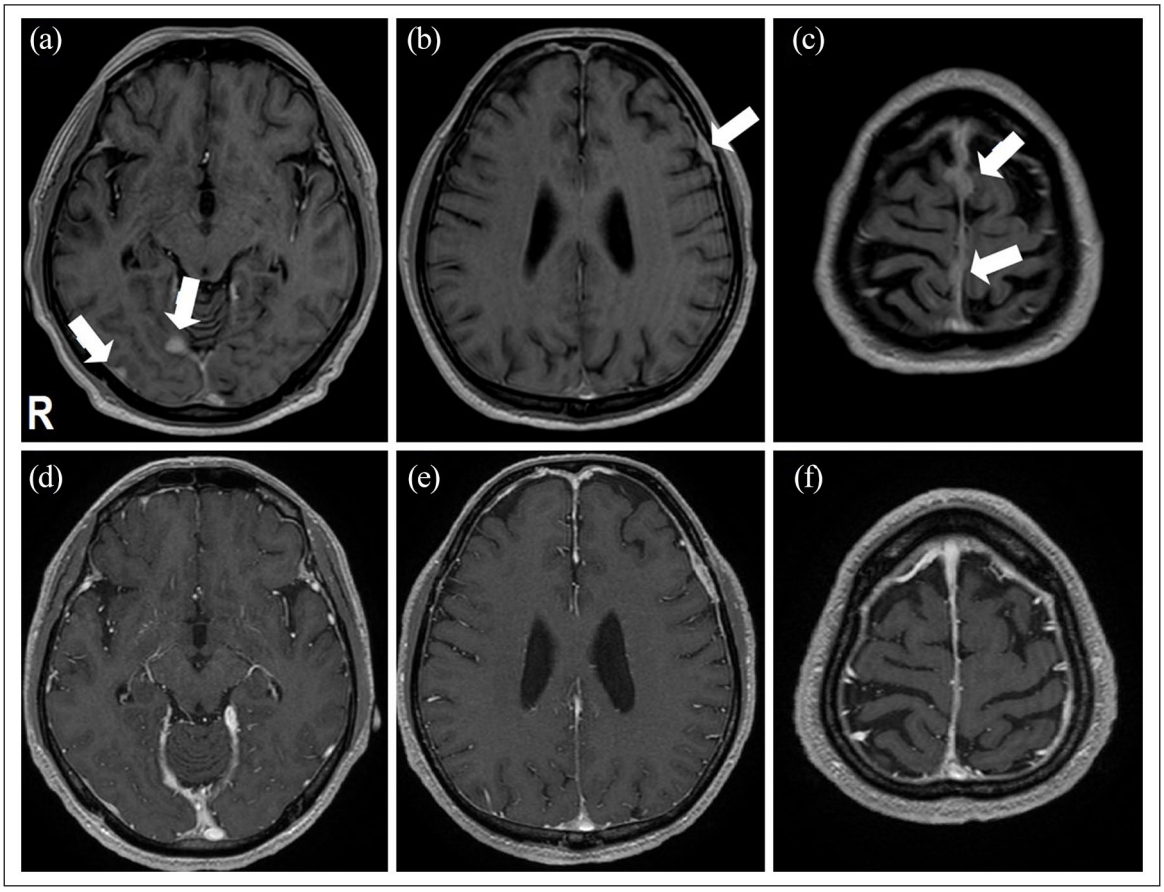

Figure 2. T1-weighted brain magnetic resonance images with contrast enhancement showing multiple dural hypertrophic lesions (arrows in a, b, and c). Follow-up neuroimaging after antituberculous treatment showing improvement of the lesions (d, e, and f).

lymphadenopathy. While cervical lymph node tuberculosis is common,[11] abdominal lymph node tuberculosis is uncommon and reported in only 0.1% of tuberculosis.[12]

Suto et al. reported a case of tuberculous myeloradiculitis and abdominal lymphadenopathy, presenting with radiculomyelopathy symptoms. Biopsy of the lymph node was culture-positive for *Mycobacterium tuberculosis*. The patient had a family history of pulmonary tuberculosis. The mechanism of infection was presumed to be a hematogenous infection from a drop infection.[13] Tsuyusaki et al. reported a case of tuberculous meningitis preceded by abdominal lymphadenopathy. Based on chest radiographic and CT findings, the patient was diagnosed with disseminated tuberculosis and showed abdominal lymphadenopathy. CSF findings also strengthened the evidence for tuberculous meningitis.[14] These rare neurological cases with abdominal lymphadenopathy exhibited some clinical evidence of tuberculosis. However, our case had no clinical history or findings of CNS tuberculosis at all before developing pachymeningitis. Since tuberculous hypertrophic pachymeningitis is rarely suspected especially if there is absent systemic disease, a biopsy is required to establish the diagnosis. Marais et al.[15] presented consensus case definition of Tuberculous meningitis. This case definition is applicable irrespective of the patient's age and HIV infection status. Extensive systemic work-up while referring to the consensus is crucial for the clinical diagnosis even in cases without positive findings of systemic tuberculosis.

The pathological mechanisms of tuberculous hypertrophic pachymeningitis and abdominal lymphadenopathy are unknown. The principal contagion method of CNS tuberculosis is infiltration into alveolar spaces. From there, secondary spread to extrapulmonary sites through bacteremia and lymphatic drainage is possible.[7] Louveau et al. demonstrated that functional lymphatic vessels line the dural sinuses. These structures express all the molecular hallmarks of lymphatic endothelial cells and can carry both fluid and immune cells from CSF and are connected to the deep cervical lymph nodes.[16] In our case, the primary focus of contagion was unclear, but we think that the abdominal paraaortic lymphatic vessel system could be involved in the dissemination of *Mycobacterium tuberculosis*.

We reported on a rare case of tuberculous hypertrophic pachymeningitis and abdominal paraaortic lymphadenopathy. Even in a case without any findings of systemic dissemination, tuberculosis should be considered a possible etiology for hypertrophic pachymeningitis.

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Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

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References

1. Tariq R and Ahmed R. Tuberculous hypertrophic pachymeningitis presenting as visual blurring and headaches. *J Pak Med Assoc* 2012; 62(9): 966–968.
2. Fonseka CL, Kanakkahewa TE, Singhapura Hewavithana JS, et al. Tuberculous pachymeningitis presenting as a diffused dural thickening in a patient with chronic headache and recurrent neurological abnormalities for more than a decade: a case report and a review of the literature. *Case Rep Infect Dis* 2018; 2018: 3012034.
3. Norbis L, Alagna R, Tortoli E, et al. Challenges and perspectives in the diagnosis of extrapulmonary tuberculosis. *Expert Rev Anti Infect Ther* 2014; 12(5): 633–647.
4. Shimoide H, Murata Y, Kusajima K, et al. The status of extrapulmonary tuberculosis in community hospital. *Kekkaku* 1994; 69(8): 519–525.
5. Mohapatra PR and Janmeja AK. Tuberculous lymphadenitis. *J Assoc Phys India* 2009; 57: 585–590.
6. Bernaerts A, Vanhoenacker FM, Parizel PM, et al. Tuberculosis of the central nervous system: overview of neuroradiological findings. *Eur Radiol* 2003; 13(8): 1876–1890.
7. Schaller MA, Wicke F, Foerch C, et al. Central nervous system tuberculosis: etiology, clinical manifestations and neuroradiological features. *Clin Neuroradiol* 2019; 29(1): 3–18.
8. Miyamoto S, Okamoto T and Nakayarna M. A case of tuberculous Jugular foramen syndrome and hypertrophic pachymeningitis. *Koutou* 2015; 27: 108–113.
9. Suárez-Calvet M, Rojas-García R, López-Contreras J, et al. Pachymeningitis, painful ophthalmoplegia, and multiple cranial neuropathy of presumed tuberculous origin. *Neuro-ophthalmology* 2011; 35(5–6): 289–292.
10. Furukawa M, Shimoda H, Kajiwara T, et al. Topographic study on nerve-associated lymphatic vessels in the murine craniofacial region by immuno-histochemistry and electron microscopy. *Biomed Res* 2008; 29(6): 289–296.
11. Ilgazli A, Boyaci H, Basyigit I, et al. Extrapulmonary tuberculosis: clinical and epidemiologic spectrum of 636 cases. *Arch Med Res* 2004; 35(5): 435–441.
12. Nonami M, Tanaka J, Azechi M, et al. Tuberculous lymphadenitis of hepatic hilar. *J Jpn Soc Intern Med* 2005; 94(10): 2189–2191.
13. Suto Y, Ito S, Nomura T, et al. A case of tuberculous myeloradiculitis with abdominal lymphadenitis presenting with symptoms of radiculomyelopathy. *Rinsho Shinkeigaku* 2015; 55(11): 816–822.
14. Tsuyusaki J, Sasaki Y, Yamagishi F, et al. Case of disseminated tuberculosis complicated with tuberculous meningitis while investigating an abdominal lymphadenopathy. *Kekkaku* 2006; 81(11): 667–671.
15. Marais S, Thwaites G, Schoeman JF, et al. Tuberculous meningitis: a uniform case definition for use in clinical research. *Lancet Infect Dis* 2010; 10(11): 803–812.
16. Louveau A, Smirnov I, Keyes TJ, et al. Structural and functional features of central nervous system lymphatics. *Nature* 2015; 523(7560): 337–341.