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Idiopathic Hypertrophic Cranial Pachymeningitis
with Accumulation of Thallium-201 on
Single-Photon Emission CT

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Summary: We report a case of idiopathic hypertrophic cranial pachymeningitis in which a high accumulation of thallium-201 was observed on an early single-photon emission CT (SPECT) scan. The patient's symptoms initially improved with steroid therapy but recurred repeatedly. MR images failed to show any change with treatment; however, thallium-201 uptake correlated closely with the fluctuation of symptoms. \(^{201}\)TI-SPECT was therefore useful in identifying inflammatory activity that was not detected by MR imaging.

Idiopathic hypertrophic cranial pachymeningitis is a rare inflammatory disease in which recurrence is frequently observed despite initial response to steroid therapy (1–4). The magnetic resonance (MR) findings in this disease are characteristic, and may be diagnostically contributory; however, they are less useful for assessing the effect of therapy, because the appearance of the lesion on MR images does not vary (4, 5). We report a patient with idiopathic hypertrophic cranial pachymeningitis in whom an accumulation of thallous chloride Tl 201 (thallium-201) on \(^{201}\)TI-single-photon emission CT (SPECT) scans correlated with fluctuation of clinical symptoms.

Case Report

A 50-year-old woman had right-sided oculomotor palsy and headache. The initial clinical diagnosis was Tolosa-Hunt syndrome. Steroid therapy was effective, and her symptoms improved. Two months later, she presented with headache and decreased visual acuity in both eyes. Results of routine laboratory tests were normal apart from a markedly elevated C-reactive protein. Serum tests for rheumatoid arthritis and syphilis, as well as anti-HIV-1 antibody, were negative. Serum angiotensin-converting enzyme level was normal, and a chest radiograph was unremarkable. Examination of CSF over the course of 1 year showed a slightly increased number of cells and protein. Contrast-enhanced CT studies showed an enhanced lesion along the right tentorial edge. MR imaging revealed a markedly thickened dural lesion of both lateral walls of the cavernous sinus that extended to the right tentorial edge (Fig 1A). The lesion appeared isointense and hypointense on T1- and T2-weighted sequences, respectively (Fig 1B), and markedly enhanced after administration of gadopentetate dimeglumine (Fig 1C). Findings at cerebral angiography were normal. Early and delayed \(^{201}\)TI-SPECT scans were obtained approximately 20 minutes and 3 hours, respectively, after intravenous injection of thallium-201. The lesion was seen to have a high accumulation of thallium-201 on the early scan, but accumulation decreased on the delayed scan (Fig 1D). Steroid therapy (methylprednisolone) was effective, and the patient's visual acuity improved in both eyes. Although the lesion appeared no different on the follow-up MR image (Fig 1E), the follow-up early \(^{201}\)TI-SPECT scan showed a decreased uptake of thallium-201 (Fig 1F).

After 2 months, the patient had a recurrence of the visual disturbance and became steroid-dependent. Over the following 7 months, her symptoms reappeared whenever treatment was tapered or discontinued. Cyclosporin and azathioprine were ineffective. A biopsy of the dural lesion was performed to rule out lymphoma or other neoplastic lesions. The specimens obtained from the right tentorial edge and anterior petroclinoid ligament showed markedly thickened, fibrotic dura with nonspecific inflammation, along with diffuse fibrotic and collagenous tissue with some lymphocytic infiltration and edema. No granulomas, multinucleated giant cells, or necrosis were present, and a diagnosis of idiopathic hypertrophic cranial pachymeningitis was made (Fig 1G). In addition to the visual disturbance, a right-sided oculomotor palsy developed, as did sensory loss of the right half of the face, despite steroid therapy. On the follow-up study, uptake of thallium-201 was again increased on the early \(^{201}\)TI-SPECT scan, but the MR imaging appearance remained unchanged. Thirteen months after the start of steroid therapy, the patient suddenly died of a retroperitoneal abscess and sepsis, both of which were probably side effects of the steroids.

Discussion

Idiopathic hypertrophic cranial pachymeningitis is a rare inflammatory disease of unknown origin that affects the meninges. The diagnosis is made by exclusion of other known causes of pachymeningitis or thickening of the dura mater, such as rheumatoid arthritis, sarcoidosis, syphilis, and tuberculosis. A review of the literature on idiopathic hypertrophic cranial pachymeningitis reveals clinical features similar to those in our case (1–6). The peak age for occurrence is in the sixth decade, and headaches and progressive cranial nerve palsies are the most common clinical features. Although these symptoms initially improve with steroid therapy, they recur or progress despite treatment in most cases (1–4). It has also been reported that the prognosis for loss of visual acuity in patients with optic nerve involvement is poor (1, 5).

The MR imaging appearance of idiopathic hypertrophic cranial pachymeningitis is characteristic and
was similar to that seen in our case. T1-weighted images typically show a markedly thickened dura with conspicuous enhancement (1–6). The lesion appears hypointense on T2-weighted images, and may have fine thin hyperintense margins (2–4, 6). It has been suggested that a central dense fibrous and collag- enous area with a more inflammatory and hypervascu- lar area at the affected dural margins may explain this typical manifestation on T2-weighted sequences (2, 3, 6). Although a dural biopsy is essential to confirm the diagnosis (2), assessment by MR imaging is helpful not only for a tentative diagnosis but also for identifying the extent of the lesion and for locating a suitable biopsy site (1). Despite improvement of symp- toms with steroid therapy, MR imaging does not show any change after steroid therapy in most cases, including ours (4, 5). Thus, follow-up MR examination may not be of value in assessing the effects of treatment and in predicting recurrence. We suggest that a marked reduction of the dense fibrotic core of the lesion is unlikely even though inflammatory activity may be suppressed by treatment.

201Tl-SPECT is widely used to detect malignant le- sions of the nervous system; however, accumulation of thallium-201 has also been reported in a few cases of cerebral infarcts and intracerebral hematomas (7–9). Furthermore, thallium-201 accumulation has been described in inflammatory lesions of various tissues.

Fig 1. 50-year-old woman with right- sided oculomotor palsy, decreased visual acuity in both eyes, and headache.

A, Axial T1-weighted noncontrast MR image (600/26 [repetition time/echo time]) shows a markedly thickened isoin- tense dural lesion (arrowheads) in both lateral walls of the cavernous sinus that extend to the right tentorial edge.

B, The lesion has a low signal inten- sity on axial T2-weighted noncontrast MR image (4000/100) (arrowheads).

C, Axial T1-weighted contrast-en- hanced MR image (600/26) shows ho- mogeneous enhancement of the lesion (arrowheads).

D, 201Tl-SPECT scan before treatment reveals an accumulation of thal- lium-201 in the lesion 20 minutes after injection (early scan, left; arrow); how- ever, accumulation decreased 3 hours after injection (delayed scan, right; ar- row). Figure continues.
Theoretically, thallium-201, which has biological properties similar to those of potassium, seems well suited as a metabolic tracer. Possible factors that may contribute to thallium-201 uptake include regional blood flow, tissue viability, sodium-potassium ATPase pump activity, and a calcium ion channel system (7–9, 12). Although it is uncertain whether an accumulation of thallium-201 in idiopathic hypertrophic cranial pachymeningitis is a common observation or not, a high uptake on an early SPECT scan that decreases on a delayed scan indicates that hypervascularity and increased lymphocytic infiltration and activity may be the most tenable explanations for the observations in the present case (7, 10, 12). Since accumulation of thallium-201 as well as the C-reactive protein value showed good correlation with fluctuation of symptoms, it was suggested that thallium-201 uptake is the best indicator of meningeal inflammation.

**Conclusion**

On the basis of the findings in this case, we propose that $^{201}$Tl-SPECT can be used to assess the effect of therapy in idiopathic hypertrophic cranial pachymeningitis and may also play a role in predicting the recurrence of this disease.
References


