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Case Report

Reversibility of CT and MR Findings in Neuro-Behçet Disease

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Abstract: A woman with neuro-Behçet disease characterized by recurrent attacks of meningoencephalitis is reported. During an attack of brain stem dysfunction characterized by dysarthria, diplopia, and ataxia, the postcontrast CT showed an enhancing lesion of the upper brain stem. During a subsequent attack characterized by subcortical dementia and amnesia, magnetic resonance (MR) showed abnormal signal intensity in the hypothalamus and upper brain stem. Upon remission of her symptoms, follow-up contrast enhanced CT and MR were entirely normal. Magnetic resonance is especially useful in localizing acute inflammatory lesions in Behçet disease. This case suggests that abnormal MR and CT findings in neuro-Behçet disease are potentially reversible and that radiographic improvement parallels clinical improvement. **Index Terms:** Behçet disease—Brain, diseases—Central nervous system, diseases—Magnetic resonance imaging—Computed tomography.

Behçet disease is an inflammatory disorder characterized by the triad of recurrent oral and genital ulcers, meningoencephalitis, and ocular inflammation. A variety of systemic manifestations may occur, including erythema nodosum, thrombophlebitis, polyarthritis, and ulcerative colitis. The CNS is involved in 25% of cases (1). Neurologic findings are variable and include meningoencephalitis, cranial nerve palsies, cerebellar ataxia, corticospinal tract dysfunction, and dementia (1–3).

The most common findings on CT in neuro-Behçet disease are low density parenchymal lesions, usually in the brain stem or basal ganglia, which show homogeneous enhancement following contrast medium infusion (4–10). Magnetic resonance (MR) imaging appears to be more sensitive than CT in detecting the brain lesions of Behçet disease (9,11–13). The CT lesions of neuro-Behçet disease appear reversible (10). The current case illustrates the reversibility of MR and CT findings in a patient with neuro-Behçet disease. The lesions seen on CT and MR correlated with the clinical deficits, and remission of symptoms was paralleled by resolution of CT and MR abnormalities.

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CASE REPORT

A 29-year-old woman was evaluated in January 1984 for a 6 month history of progressive neurologic dysfunction that included blurred vision, diplopia, and headaches. Two months prior to admission, she experienced episodes of dysarthria. Subsequently, she noted generalized malaise, occasional light-headedness, and weakness of the left arm and leg. On admission, neurologic examination demonstrated dysarthria, bilateral internuclear ophthalmoplegia, bilateral ataxia, bilateral dysdiadochokinesia, and a wide-based gait. Ulcerated lesions were noted on examination of the oral mucosa. Cerebrospinal fluid examination showed 121 white blood cells/mm^3 (74% lymphocytes) and protein elevated to 60 mg/dl. Contrast enhanced CT demonstrated an enhancing midbrain lesion (Figs. 1a and b). High dose corticosteroids were administered and her symptoms resolved.

Five months later the corticosteroids were discontinued. One month later the patient experienced recurrent headaches, high fever, and ulcers of the vulvar mucosa. Vulvar biopsy showed nonspecific inflammation. Contrast enhanced CT at this time was normal. Based on

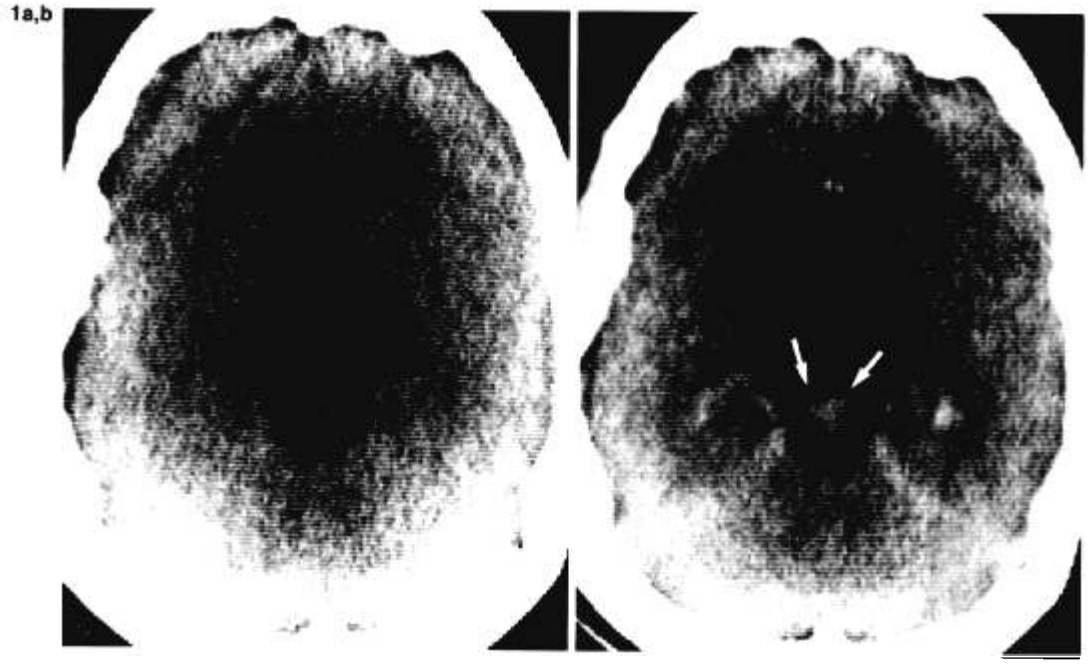

FIG. 1. a: Plain CT is normal. **b:** Postcontrast CT shows enhancement in the midbrain (arrows). Patient had acute attack of meningoencephalitis characterized by lethargy, diplopia, truncal ataxia, and dysarthria.

recurrent oral and genital ulcers associated with recurrent meningoencephalitis, a diagnosis of neuro-Behçet disease was made.

Over the ensuing 3 years, the patient visited the emergency room frequently with recurrent headaches, high fever, and ulcers of the genitalia and oral mucosa.

In September 1987, she experienced an exacerbation of her disease characterized by high fever, memory impair-

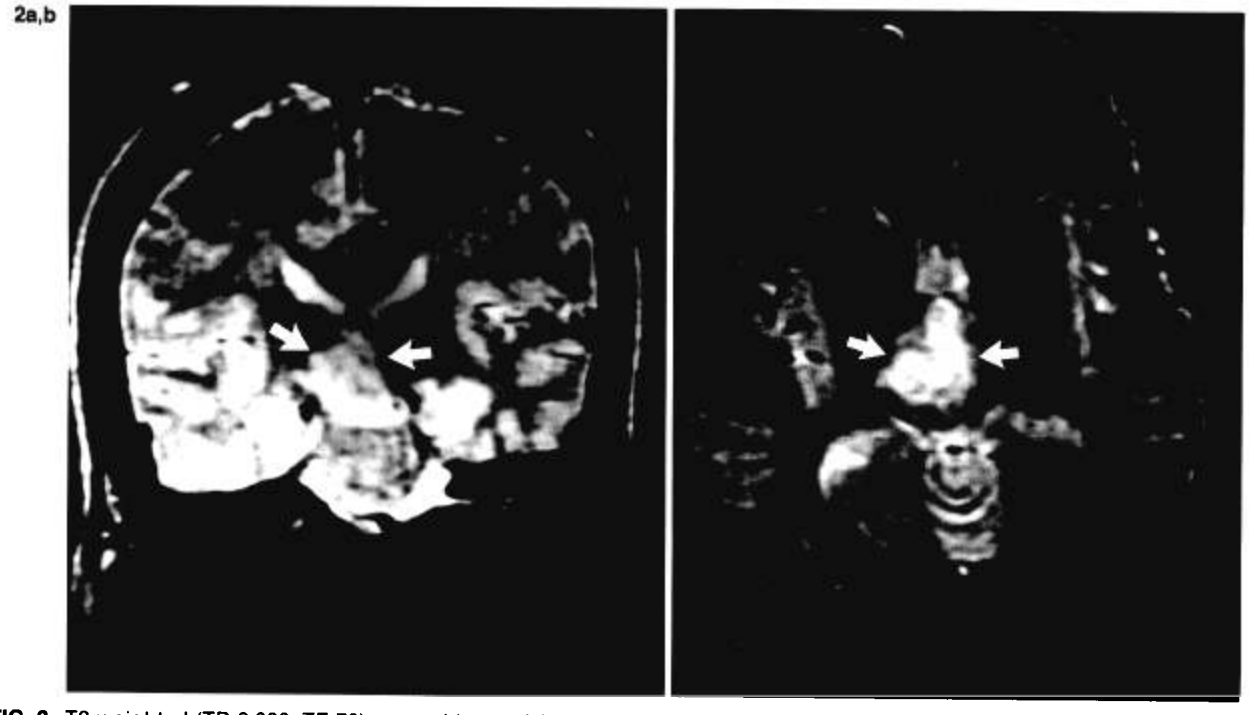

FIG. 2. T2-weighted (TR 2,000, TE 70) coronal image **(a)** axial image **(b)** demonstrate high signal lesion involving midbrain and thalami on both sides (arrows), more on the right than left. Patient was experiencing subacute attack of meningoencephalitis characterized by dementia and amnesia.

ment, dementia, and headaches. There were no focal neurologic signs. An MR evaluation was obtained with multiple spin echo sequences using T1 [repetition time (TR)600, echo time (TE)25], T2 (TR 2,000, TE 70) (Figs. 2a, 2b), and proton density (TR 2,000, TE 20) weighting in multiple planes. Discrete areas of abnormal high signal intensity, most prominent in T2-weighted images, appeared in the midbrain and hypothalamus. The headache and fever resolved with high dose corticosteroid therapy. However, the memory impairment and dementia persisted.

Three weeks later the patient experienced yet another exacerbation, presenting as pseudobulbar weakness including difficulties in swallowing, headache, high fever, and lethargy. Plain CT showed areas of low attenuation in the right basal ganglia and left thalamus. On contrast enhanced CT, the thalamic lesion showed contrast enhancement (Fig. 3). There was no correlate on CT to the midbrain lesion previously visualized by MR. Lethargy and brain stem symptoms resolved after resumption of corticosteroids. For the next 12 months she was treated concurrently with low dose corticosteroids and azathioprine. At that time she was clinically asymptomatic. Contrast enhanced CT as well as gadolinium enhanced MR were completely normal (Figs. 4 and 5). The patient remains asymptomatic on combination therapy with prednisone and azathioprine.

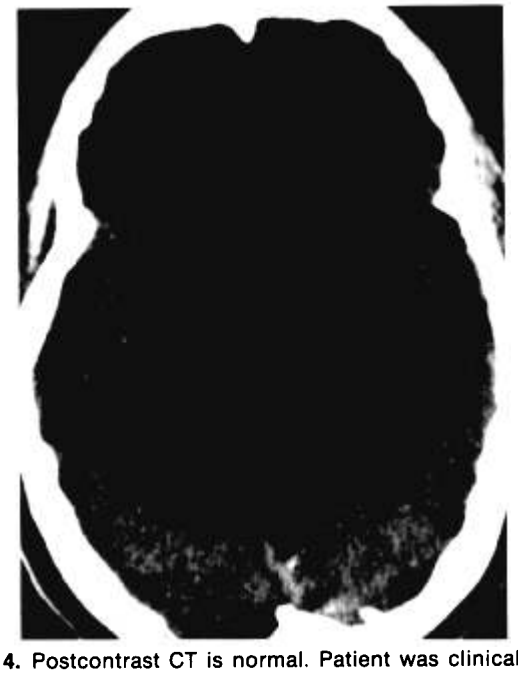

FIG. 4. Postcontrast CT is normal. Patient was clinically in remission.

DISCUSSION

Behçet disease, originally described as the triad of aphthous stomatitis, genital ulceration, and uveitis (14), is now recognized to encompass both systemic and neurologic manifestations. Although controversy over diagnostic criteria remains, most authorities require a combination of major criteria (oral ulcers, genital ulcers, ocular inflammation) with or without minor criteria (neurologic, gastrointestinal, vascular, and joint involvement) (15,16). Onset is generally in the third decade; men are af-

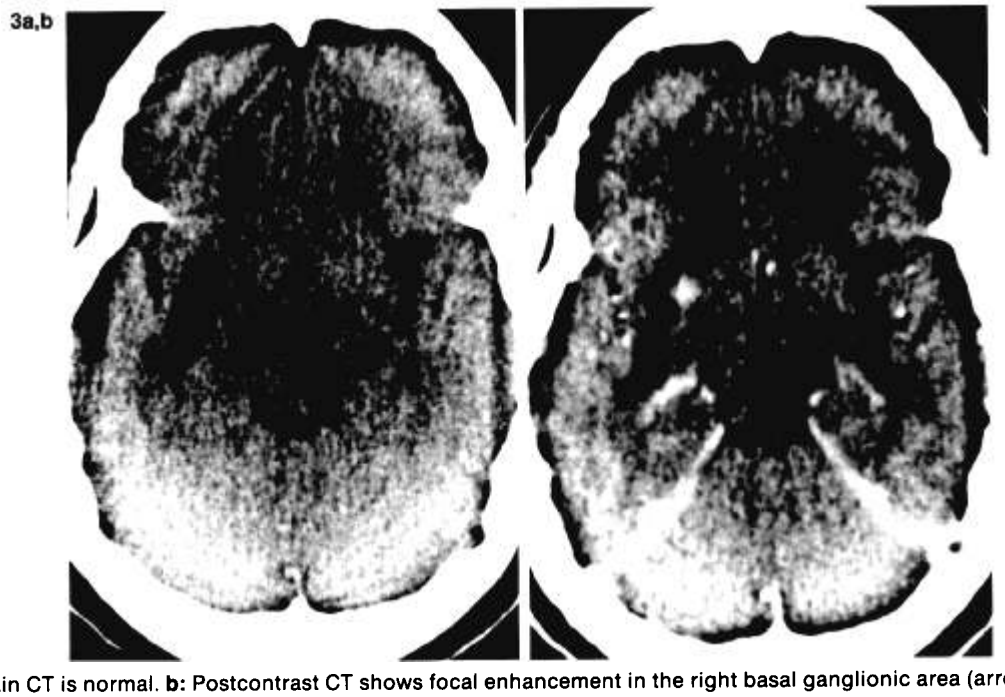

FIG. 3. a: Plain CT is normal. **b:** Postcontrast CT shows focal enhancement in the right basal ganglionic area (arrows). Midbrain and thalami are normal.

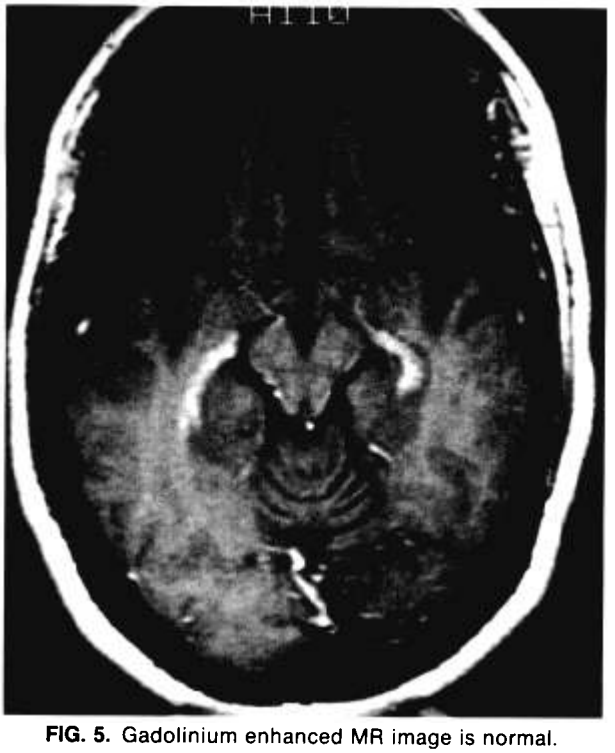

FIG. 5. Gadolinium enhanced MR image is normal.

fected about twice as frequently as women. Although the etiology is unknown, support exists for both viral and autoimmune mechanisms (16). Histologically, a vasculitic picture with lymphocytic infiltration predominates (2,17).

Neurologic involvement in Behçet disease occurs in 10–25% of cases (1) and is the presenting manifestation in up to 5% of cases. Central nervous system involvement includes intracranial hypertension secondary to dural sinus thrombosis, intracranial inflammatory lesions with predilection for deep structures (primarily brain stem and basal ganglia), and recurrent meningoencephalitis (18,19). Our patient manifested both recurrent aseptic meningitis as well as a subcortical dementia (20) related to inflammatory lesions of the basal ganglia and brain stem.

In eight previously reported cases of neuro-Behçet disease, all but one showed MR abnormalities (9,11–13). Lesions were more likely to occur in the brain stem, basal ganglia, thalamus, and internal capsule. When MR has been compared to CT, MR has been found to better delineate lesions seen on CT as well as to show some lesions not seen on CT. Magnetic resonance is especially superior to CT in the ventral brain stem region where CT is hindered by bone hardening artifacts. Long-term follow-up was not reported for these cases. Therefore, it was not possible to determine whether these lesions resolve or progress to chronic gliosis. Our case illustrates that clinical remission may be associated with complete MR and CT scan resolution of inflammatory lesions.

In the current case, the MR and CT images showed good correlation with the patient's clinical state. When the patient presented initially with a brain stem syndrome of ataxia and dysarthria, the CT demonstrated an abnormal area in the upper brain stem. Later, when the patient showed signs of subcortical dementia with memory impairment, the MR and CT findings demonstrated abnormal areas of signal in the thalamus and basal ganglia. Finally, after treatment, remission of clinical symptoms was associated with a resolution of all abnormal signals on MR and CT. We conclude that MR and CT abnormalities in neuro-Behçet disease are potentially reversible. This conclusion is consistent with the suggestion of Herskovitz et al. (10) that CT lesions in neuro-Behçet disease are reversible and that improvement in these lesions parallels clinical improvement. Herskovitz et al. suggest that the CT lesion in neuro-Behçet disease may reflect a reversible breakdown in the blood–brain barrier (possibly related to inflammation) rather than gliosis or infarction. The reversibility of MR findings in our case is consistent with this hypothesis

REFERENCES

1. Wolf SM, Schotland DL, Phillips LL. Involvement of nervous system in Behçet's syndrome. *Arch Neurol* 1965;12: 315–25.
2. Chajek T, Fainaru M. Behçet's disease. Report of 41 cases and a review of the literature. *Medicine* 1975;54:179–96.
3. O'Duffy JD, Goldstein NP. Neurologic involvement in seven patients with Behçet's disease. *Am J Med* 1976;61:170–8.
4. Kozin F, Haughton V, Bernhard GC. Neuro-Behçet disease: two cases and neuroradiologic findings. *Neurology* 1977;27:1148–52.
5. Williams AL, Haughton VM, Saxena VK, Albers JW. Computed tomography in Behçet's disease. *Radiology* 1979;131:403–4.
6. Dobkin BH. Computerized tomographic findings in neuro-Behçet's disease. *Arch Neurol* 1980;37:58–9.
7. Kuroiwa H, Tohgi H, Kanayama H, Fujimori M, Aoki H. Neuro-Behçet's disease with alternating hemiparesis. *Neuroradiology* 1986;28:284.
8. Weitz Z, Machtey I, Rothman GM, Davidovich S, Schifter T. Computed axial tomography of brain in neuro-ocular Behçet's syndrome. *Z Rheumatol* 1986;45:310–1.
9. Willeit J, Schmutzhard E, Aichner F, Mayr U, Weber F, Gerstenbrand F. CT and MR imaging in neuro-Behçet disease. *J Comput Assist Tomogr* 1986;10:313–5.
10. Herskovitz S, Lipton RB, Lantos G. Neuro-Behçet's disease: CT and clinical correlates. *Neurology* 1988;38:1714–20.
11. Fukuyama H, Kameyama M, Nabatame H, et al. Magnetic resonance images of neuro-Behçet syndrome show precise brain stem lesions: report of a case. *Acta Neurol Scand* 1987;75:70–3.
12. Kojma S, Fukutake T, Hirayama K, Iwamoto I. Magnetic resonance imaging in neuro-Behçet's disease. *Rinsho Shinkeigaku* 1987;27:458–64.
13. Miller DH, Ormerod IEC, Gibson A, du Boulay EPGH, Rudge P, McDonald WI. MR brain scanning in patients with

vasculitis: differentiation from multiple sclerosis. *Neuroradiology* 1987;29:226–31.
14. Behçet H. Uber rezidivierende, aphthose, durch ein Virus verursachte Geschwure am Mund, am Auge und an den Genitalien. *Dermatol Wochenschr* 1937;105:1152–7.
15. Ammann AJ, Johnson A, Fyfe GA, Leonards R, Wara DW, Cowan MJ. Behçet syndrome. *J Pediatr* 1985;107:41–3.
16. Jorizzo JL. Behçet's disease: an update based on the 1985 International Conference in London. *Arch Dermatol* 1986;122:556–8.
17. James DG. Silk route disease. *Postgrad Med J* 1986;62: 151–3.
18. Davis LE, Hodgin UG, Kornfeld M. Recurrent meningoencephalitis with recovery from Behçet's disease. *West J Med* 1986;145:238–9.
19. Litvan I, Roig C, Rovira A, Ruscalleda J. Behçet's syndrome masquerading as tumor. *Neuroradiology* 1987;29:103.
20. Cummings JL, Benson DF. Subcortical dementia: review of an emerging concept. *Arch Neurol* 1984;41:874–9.