

5. De Michele G, Maltecca F, Carella M, et al. Dementia, ataxia, extrapyramidal features, and epilepsy: phenotype spectrum in two Italian families with spinocerebellar ataxia type 17. *Neurol Sci* 2003;24:166–167.
6. Fujigasaki H, Martin JJ, De Deyn PP, et al. CAG repeat expansion in the TATA box-binding protein gene causes autosomal dominant cerebellar ataxia. *Brain* 2001;24:1939–1947.
7. Gunther P, Storch A, Schwarz J, et al. Basal ganglia involvement of a patient with SCA 17: a new form of autosomal dominant spinocerebellar ataxia. *J Neurol* 2004;251:896–897.
8. Hagenah JM, Zuhlke C, Hellenbroich Y, Heide W, Klein C. Focal dystonia as a presenting sign of spinocerebellar ataxia 17. *Mov Disord* 2004;19:217–220.
9. Maltecca F, Filla A, Castaldo I, et al. Intergenerational instability and marked anticipation in SCA-17. *Neurology* 2003;61:1441–1443.
10. Nakamura K, Jeong SY, Uchiyama T, et al. SCA17, a novel autosomal dominant cerebellar ataxia caused by an expanded polyglutamine in TATA-binding protein. *Hum Mol Genet* 2001;10:1441–1448.
11. Oda M, Maruyama H, Komure O, et al. Possible reduced penetrance of expansion of 44 to 47 CAG/CAA repeats in the TATA-binding protein gene in spinocerebellar ataxia type 17. *Arch Neurol* 2004;61:209–212.
12. Rolfs A, Koeppen AH, Bauer I, et al. Clinical features and neuropathology of autosomal dominant spinocerebellar ataxia (SCA17). *Ann Neurol* 2003;54:367–375.
13. Silveira I, Miranda C, Guimaraes L, et al. Trinucleotide repeats in 202 families with ataxia: a small expanded (CAG)_n allele at the SCA17 locus. *Arch Neurol* 2002;59:623–629.
14. Stevanin G, Fujigasaki H, Lebre AS, et al. Huntington's disease-like phenotype due to trinucleotide repeat expansions in the TBP and JPH3 genes. *Brain* 2003;126:1599–1603.
15. Toyoshima Y, Yamada M, Onodera O, et al. SCA17 homozygote showing Huntington's disease-like phenotype. *Ann Neurol* 2004;55:281–286.
16. Zuhlke C, Hellenbroich Y, Dalski A, et al. Different types of repeat expansion in the TATA-binding protein gene are associated with a new form of inherited ataxia. *Eur J Hum Genet* 2001;9:160–164.
17. Zuhlke C, Gehlken U, Hellenbroich Y, Schwinger E, Burk K. Phenotypical variability of expanded alleles in the TATA-binding protein gene. Reduced penetrance in SCA17? *J Neurol* 2003;250:161–163.
18. Zuhlke CH, Spranger M, Spranger S, et al. SCA17 caused by homozygous repeat expansion in TBP due to partial isodisomy 6. *Eur J Hum Genet* 2003;11:629–632.
19. Gostout B, Liu Q, Sommer SS. "Cryptic" repeating triplets of purines and pyrimidines (cRRY(i)) are frequent and polymorphic: analysis of coding cRRY(i) in the proopiomelanocortin (POMC) and TATA-binding protein (TBP) genes. *Am J Hum Genet* 1993;52:1182–1190.
20. Duyao M, Ambrose C, Myers R, et al. Trinucleotide repeat length instability and age of onset in Huntington's disease. *Nat Genet* 1993;4:387–392.
21. Payami H, Nutt J, Ganther S, et al. SCA2 may present as levodopa-responsive parkinsonism. *Mov Disord* 2003;18:425–429.
22. Imamura A, Sugai K, Watanabe S, Hamada F, Kurashige T, Takashima S. High intensity in the globus pallidus on proton and T2-weighted MRI in a case of dentato-rubro-pallido-luysian atrophy of myoclonus epilepsy type. *Acta Paediatr Jpn* 1994;36:527–530.
23. Tanaka M, Hirai S, Kondo S, et al. Cerebral hypoperfusion and hypometabolism with altered striatal signal intensity in chorea-acanthocytosis: a combined PET and MRI study. *Mov Disord* 1998;13:100–107.
24. Wray SH, Provenzale JM, Johns DR, Thulborn KR. MR of the brain in mitochondrial myopathy. *AJNR Am J Neuroradiol* 1995;16:1167–1173.
25. Schrag A, Kingsley D, Phatouros C, et al. Clinical usefulness of magnetic resonance imaging in multiple system atrophy. *J Neurol Neurosurg Psychiatry* 1998;65:65–71.

Paraneoplastic Chorea With Leukoencephalopathy Presenting With Obsessive–Compulsive and Behavioral Disorder

Susanne Muehlschlegel, MD,^{1*} Michael S. Okun, MD,^{1,2}
Kelly D. Foote, MD,² Dominique Coco, MD,³
Anthony T. Yachnis, MD,³ and Hubert H. Fernandez, MD¹

¹*Department of Neurology, Movement Disorders Center,
University of Florida College of Medicine, Gainesville,
Florida, USA*

²*Department of Neurological Surgery, University of Florida
College of Medicine, Gainesville, Florida, USA*

³*Department of Pathology, Immunology and Laboratory
Medicine, University of Florida College of Medicine,
Gainesville, Florida, USA*

Abstract: Chorea is a rare manifestation of paraneoplastic disease and is associated with CV2/CRMP-5 antibodies. Obsessive–compulsive disorder and large-scale white matter abnormalities on MRI have not been previously reported in association with these antibodies. We report on a case of CV2 paraneoplastic syndrome with obsessive–compulsive behavior preceding the motor manifestations of chorea with associated leukoencephalopathy on MRI. The literature on paraneoplastic chorea is reviewed. © 2005 Movement Disorder Society

Key words: CV2 paraneoplastic antibodies; paraneoplastic chorea; obsessive–compulsive disorder; leukoencephalopathy



Progressive chorea is a rare presentation of paraneoplastic central nervous system disease. It has been reported with small cell lung cancer,^{1–7} thymoma,⁸ lymphoma,^{9,10} and renal cell carcinoma.¹¹ The associated paraneoplastic antibodies in these cases are CRMP-5/CV2.^{8,12} We describe the clinicopathological and radiological findings of a patient with CV2 serum antibodies whose initial presentation mimicked many of the features of Huntington's disease with personality changes and obsessive–compulsive behavior prior to development of severe chorea.

This article contains Supplementary Video, available online at www.interscience.wiley.com/jpages/0885-3185/suppmat.

*Correspondence to: Dr. Susanne Muehlschlegel, PO Box 100236, Gainesville, FL 32605. E-mail: muehlschlegel@neurology.ufl.edu

Received 10 October 2004; Revised 21 January 2005; Accepted 26 January 2005

Published online 21 July 2005 in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/mds.20570

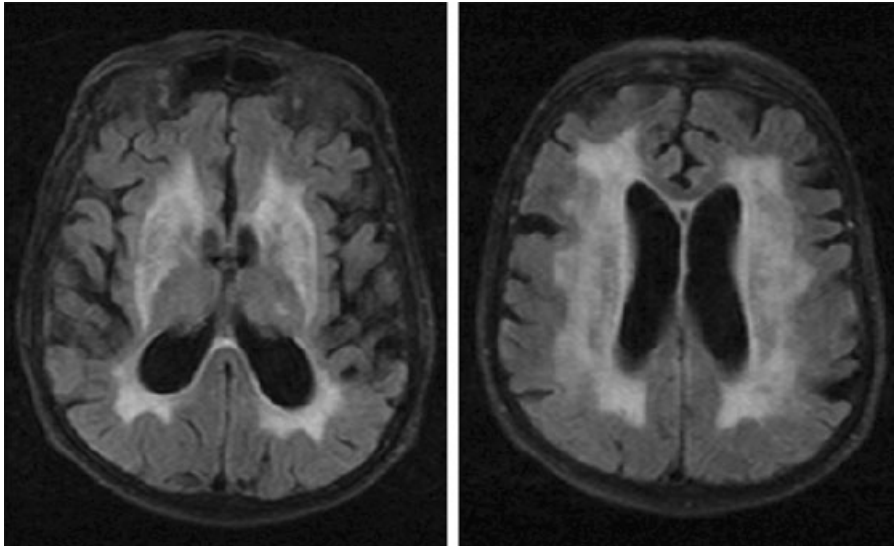


FIG. 1. FLAIR images of axial brain MRI with T2 hyperintense signal changes of the subcortical white matter, striatum, globus pallidus sparing the thalami bilaterally.

Case Report

A 69-year-old woman with no significant medical history but a more than 60-pack-year smoking history was noted to develop irrational and argumentative behavior in September 2003. Obsessive-compulsive behavior developed within the first month of her illness. She placed objects in distinct lined-up order on every bed and table in the house and was constantly rearranging the order. Four weeks later, her family noticed development of severe choreic symptoms and unsteady gait. Workup at two different local hospitals included a brain MRI revealing white matter changes. MRA showed 90% stenosis of the right ICA and a right carotid endarterectomy was performed with no effect on her symptoms. Gradual progression of her movement disorder was noted. She was medicated with alprazolam, haloperidol, levetiracetam, and lorazepam without benefit. Serum ceruloplasmin, copper, anti-streptolysin-O-titer, B12, thyroid stimulating hormone, liver function tests, rheumatoid factor, erythrocyte sedimentation rate, anti-nuclear antibody, C3- and C4-complement, and homocysteine levels were checked and found to be normal. A repeat brain MRI 6 months into her illness showed severe leukoencephalopathy with symmetrical deep and subcortical white matter hyperintense T2 signal changes also involving the striatum and globus pallidus but sparing the thalami bilaterally (Fig. 1). There was some cortical atrophy and mild atrophy of the head of the caudate (without contrast enhancement). She was diagnosed with tardive dyskinesia. All medications except alprazolam were stopped. She was treated with B6 and melatonin without improvement.

Seven months into her illness she was no longer able to ambulate and exhibited dysarthria as well as urinary and bowel incontinence. She was referred to our institution for a second opinion. There had been weight loss of 40 lb over the previous 7 months. She was unable to communicate verbally and was unable to eat or drink due to her involuntary movements. Neurological examination revealed severe dysarthria with widely varying intonation of her voice (choreic vs. ataxic) (see Video, Segment 1). She could only communicate by showing thumbs up or down. She was following simple commands, but orientation, attention, and higher cortical functions could not be adequately assessed. She had slow horizontal and vertical sac-

ades bilaterally. Motor examination revealed paratonia with severe chorea and biballism involving the upper greater than lower extremities, with some mild finger dystonia symmetrically. She had frequent tongue protrusion, facial grimacing, eyelid blinking, and eyebrow raising with head twisting (see Video, Segments 2 and 3). Deep tendon reflexes were 2+ throughout, including ankles. She had bilateral striatal toes with a plantar flexor response on the right and extensor on the left. Sensory testing, limited to nail bed pressure, was symmetric. She was unable to sit or stand and required high bed rails to avoid falling out of bed.

The following additional laboratory tests were normal: complete blood count, blood smear looking for acanthocytes, coagulation panel, basic metabolic panel, magnesium, phosphorus, thyroid peroxidase antibodies, thyroglobulin antibodies, antiphospholipid antibodies, Huntington's disease gene test, and Rickettsia serum antibodies. Cerebrospinal fluid (CSF) examination revealed four white blood cells/mm³ (no differential was performed) with 48 mg/dl protein. CSF glucose, cytology, cultures (bacterial, viral, fungal, and acid fast bacteria), cryptococcal antigen, VDRL, protein 14-3-3, lyme titer, and West Nile virus antibodies were normal or negative. A full serum paraneoplastic antibody panel (Athena Diagnostics) was sent and CV2 antibodies were reported as positive with presence of a 37 kD band on Western blot using purified human recombinant antigen. The positive Western blot was confirmed by immunohistochemical analysis in the laboratory of Dr. Jerome Honnorat in Lyon, France.

MRI was repeated 3 weeks after admission and was unchanged from the prior MRI. A chest X-ray suggested an increased opacity in the medial aspect of the left apex; however, an apical lordotic view did not confirm this finding. A chest CT was not performed, as cancer was initially not suspected. EEG interpretation was limited due to the constant movements but showed left focal slowing without periodic discharges.

A right frontal brain biopsy including meninges, cortex, and deep white matter was performed. It revealed cerebral amyloid angiopathy and scanty perivascular infiltrates of mature T lym-

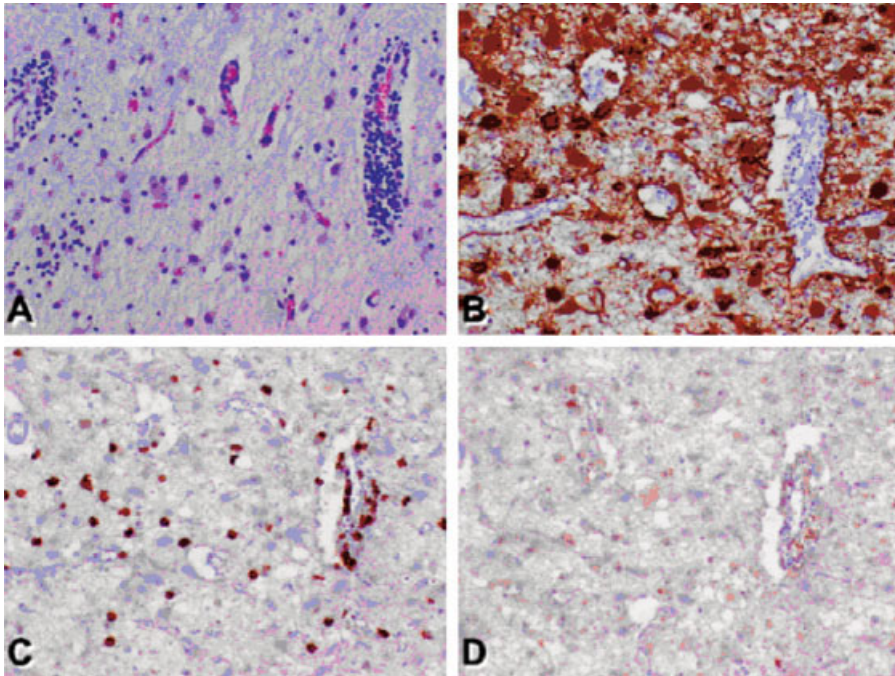


FIG. 2. **A:** H&E-stained section from the caudate nucleus showing perivascular and parenchymal infiltrates of mature lymphocytes, diffuse neuronal loss, and reactive (hypertrophic) gliosis. **B:** GFAP-immunostained section showing reactive gliosis. **C:** CD8-immunostained section showing strong reactivity consistent with cytotoxic/suppressor T lymphocytes. **D:** CD4 (T-helper cell)-immunostained section showing only rare positivity. Original magnification = 500 \times . [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

phocytes. Nonspecific gliosis was also noted. The patient was treated with 3 days of high-dose steroids without improvement. Cyclophosphamide treatment was offered but declined by the family. She died 5 weeks later.

Autopsy Findings

Consent for autopsy was limited to the brain. The unfixed brain weighed 1,200 gm. The leptomeninges were clear and there was mild sulcal widening of the surface convexities. Serial section of the cerebral hemispheres revealed dusky discoloration of the basal ganglia, which was most prominent in the striatum. The caudate nucleus was slightly reduced in volume and had a granular gut surface.

The most striking microscopic findings were extensive neuronal loss, reactive astrocytosis, and perivascular and parenchymal infiltrates of mature lymphocytes (Fig. 2A) limited to the striatum. Immunohistochemical studies confirmed a brisk reactive gliosis, which was composed of hypertrophic glial fibrillary acidic protein (GFAP)-positive astrocytes (Fig. 2B). The parenchymal lymphocytic infiltrate in the striatum was composed exclusively of CD8-immunoreactive T lymphocytes consistent with T cytotoxic/suppressor cells (Fig. 2C,D), while perivascular infiltrates contained both B and T lymphocytes (CD4- and CD8-reactive). This pattern of T-cell inflammation has been well documented in paraneoplastic encephalitis.¹³

Arterial blood vessels of the subcortical and deep white matter and basal ganglia showed widespread hyalinization and arteriosclerosis consistent with hypertensive vascular change. Cerebral amyloid angiopathy was confirmed by immunohistochemical staining for A- β -amyloid peptide. There were focal β -amyloid-positive neuritic plaques in cortical association areas, but not sufficient to satisfy criteria for Alzheimer's disease. Immunostains for τ , α -synuclein and ubiquitin performed on several levels of the neuraxis were all negative. There were no

areas of demyelination seen to suggest progressive multifocal leukoencephalopathy (PML) as seen by JC virus infection.

Unfortunately in the case of our patient, the CV2 antibodies were reported positive several days after her death. All other paraneoplastic antibodies, including anti-Hu, were negative. Autopsy consent was given only for the brain so that the primary malignancy could not be identified.

Discussion

Paraneoplastic CNS disease is believed to be the result of an antigen-driven oligoclonal cytotoxic T-cell response.¹³ The pattern of T-cell infiltration, as seen in our case, is the pathological signature of paraneoplastic disease. In combination with the CV2 antibodies, we presumed the clinical syndrome (behavioral and motor) to be the result of paraneoplastic disease.

Although the first case of paraneoplastic chorea was reported in 1988,¹ CV2 paraneoplastic antibodies were first described in 1996¹² and initially linked only to peripheral neuropathy, ataxia, uveitis, and optic neuritis.^{12,14,15} CRMP-5 antibodies were described in 2001 and have been reported more frequently with paraneoplastic chorea and limbic encephalitis but also with visual loss and cranial nerve palsies.^{5,8} It has been proposed that both CRMP-5 and CV2 are the same antibody or group of antibodies.¹⁶ Until today, paraneoplastic chorea has been reported in a total of 27 to 38 patients; two reports were case series, which is why some cases may be duplicates counted in both reports. Other neurological manifestations, some in association with simultaneous anti-Hu (ANNA-1) antibody, include cranial nerve palsies, vision loss, limbic encephalitis, and dystonia.^{1-11,17} The most interesting finding in this case was the effect on nonmotor basal ganglia circuitry with obsessive-compulsive features, which have not been described in paraneoplastic chorea. The presence of these abnormalities early in the neurological picture suggests a different

presentation of paraneoplastic syndromes (nonmotor before motor features). Similar to Huntington's disease and obsessive-compulsive disorder (OCD), we propose that nonmotor loops of basal ganglia, as described previously,¹⁸ and pathways in the family of parallel circuits involving the anterior cingulate, dorsolateral prefrontal cortex, and orbitofrontal cortex are affected and responsible for the nonmotor features present in this case. The involvement of these structures in OCD, but also the direct involvement of the basal ganglia, is supported by structural MRI studies and three-dimensional volumetric imaging studies in OCD with demonstration of increased gray matter volume in the ventral putamen¹⁹ and decreased globus pallidus volumes.²⁰ A SPECT study shows altered dopamine transporter density in the basal ganglia in patients with OCD suggesting an association of the dopaminergic neurotransmitter system in this disease.²¹ Involvement of the oculomotor loop likely resulted in the slow saccades as it does in Huntington's disease. Interestingly, oculomotor, motor, and dorsolateral prefrontal loops seemed to be all involved. This diffuse involvement suggests that the CV2 antibody syndrome started predominantly with nonmotor loops, but did not spare any of the basal ganglia circuitry. The reasons for this antibody's predilection for these brain regions remain to be elucidated. There was no pathological correlate to the clinical findings seen in the brainstem during autopsy suggesting localization of the oculomotor and motor signs above the level of the brainstem.

On review of the previously reported cases of paraneoplastic chorea, MRI findings seemed to be independent from clinical presentation. Most previous cases showed MRI involvement of the caudate nucleus and pallidum with sparing of the thalamus, or patchy basal ganglia involvement.^{3-5,7,9} Paraneoplastic basal ganglia degeneration with widespread leukoencephalopathy changes as seen in our patient has not previously been noted. Likely etiologies for the subcortical T2 signal changes seen on MRI include paraneoplastic changes, extensive small-vessel cerebrovascular disease, and cerebral amyloid angiopathy (CAA). Given that our patient had some risk factors for small-vessel disease, including smoking and hypertension, it is possible that the T2 white matter changes were independent from paraneoplastic disease and amyloid angiopathy and due to atherosclerotic changes in the white matter found on autopsy. Reports in the literature suggest that CAA can be associated with inflammation²² or may even coexist with primary CNS vasculitis.²³ The brain biopsy was suggestive of such, which is why immunosuppressive therapy was started, though without immediate clinical response. The autopsy, however, did not confirm CNS vasculitis or inflammation other than described with paraneoplastic disease. No foci of demyelination were seen, which makes PML highly unlikely.

As a criticism in this case, a chest CT or whole PET scan would have been helpful investigations in the search for a primary malignancy but were not undertaken because cancer was not highly suspected in this patient. Also, antibasal ganglia antibodies were not searched, although they can have a possible diagnostic utility in the evaluation of idiopathic movement disorders.²⁴

This unique case underlines the importance of considering paraneoplastic chorea in cases of nonmotor as well as progressive unexplained motor manifestations of hyperkinetic movement disorders. Preceding symptoms involving behavior should be examined carefully. Leukoencephalopathy may be more than patchy and can be seen on MRI in addition to striatal T2 signal abnormalities. Early testing for CRMP-5/CV2 antibodies is very important and should be performed in all cases

of unexplained chorea. Although pharmacological treatment may provide symptomatic improvement in some cases, the mainstay of therapy is early detection and treatment of the underlying malignancy.

Legends to the Video

Segment 1. The patient displays significant facial and voice chorea.

Segment 2. The patient has chorea and biballism in bilateral upper extremities, face and neck with some finger dystonia and striatal toes.

Segment 3. The patient has slow saccades. We are holding the patient's head as she was unable to suppress lateral head movement with the saccades.

Acknowledgments: We thank Mr. Charles Jacobson for the technical support with the video material.

References

- Albin RL, Bromberg MB, Penney JB, Knapp R. Chorea and dystonia: a remote effect of carcinoma. *Mov Disord* 1988;3:162-169.
- Heckmann JG, Lang CJ, Druschky A, Claus D, Bartels O, Neundorfer B. Chorea resulting from paraneoplastic encephalitis. *Mov Disord* 1997;12:464-466.
- Tani T, Piao Y, Mori S, et al. Chorea resulting from paraneoplastic striatal encephalitis. *J Neurol Neurosurg Psychiatry* 2000;69:512-515.
- Croteau D, Owainati A, Dalmau J, Rogers LR. Response to cancer therapy in a patient with a paraneoplastic choreiform disorder. *Neurology* 2001;57:719-722.
- Vernino S, Tuite P, Adler CH, et al. Paraneoplastic chorea associated with CRMP-5 neuronal antibody and lung carcinoma. *Ann Neurol* 2002;51:625-630.
- Tremont-Lukats IW, Fuller GN, Ribalta T, Giglio P, Groves MD. Paraneoplastic chorea: case study with autopsy confirmation. *Neuro-oncology* 2002;4:192-195.
- Kinirons P, Fulton A, Keoghan M, Brennan P, Farrell MA, Moroney JT. Paraneoplastic limbic encephalitis (PLE) and chorea associated with CRMP-5 neuronal antibody. *Neurology* 2003;61:1623-1624.
- Yu Z, Kryzer TJ, Griesmann GE, Kim K, Benarroch EE, Lennon VA. CRMP-5 neuronal autoantibody: marker of lung cancer and thymoma-related autoimmunity. *Ann Neurol* 2001;49:146-154.
- Batchelor TT, Platten M, Palmer-Toy DE, et al. Chorea as a paraneoplastic complication of Hodgkin's disease. *J Neurooncol* 1998;36:185-190.
- Nuti A, Ceravolo R, Salvetti S, Gambaccini G, Bonuccelli U, Capochiani E. Paraneoplastic choreic syndrome during non-Hodgkin's lymphoma. *Mov Disord* 2000;15:350-352.
- Kujawa KA, Niemi VR, Tomasi MA, Mayer NW, Cochran E, Goetz CG. Ballistic-choreic movements as the presenting feature of renal cancer. *Arch Neurol* 2001;58:1133-1135.
- Honnorat J, Antoine JC, Derrington E, Aguera M, Belin MF. Antibodies to a subpopulation of glial cells and a 66 kDa developmental protein in patients with paraneoplastic neurological syndromes. *J Neurol Neurosurg Psychiatry* 1996;61:270-278.
- Voltz R, Dalmau J, Posner JB, Rosenfeld MR. T-cell receptor analysis in anti-Hu associated paraneoplastic encephalomyelitis. *Neurology* 1998;51:1146-1150.
- Antoine JC, Honnorat J, Camdessanche JP, et al. Paraneoplastic anti-CV2 antibodies react with peripheral nerve and are associated with a mixed axonal and demyelinating peripheral neuropathy. *Ann Neurol* 2001;49:214-221.
- de la Sayette V, Bertran F, Honnorat J, Schaeffer S, Iglesias S, Defer G. Paraneoplastic cerebellar syndrome and optic neuritis with anti-CV2 antibodies: clinical response to excision of the primary tumor. *Arch Neurol* 1998;55:405-408.

16. Honnorat J, Antoine JC, Belin MF. Are the "newly discovered" paraneoplastic anticollapsin response-mediator protein 5 antibodies simply anti-CV2 antibodies? *Ann Neurol* 2001;50:688–691.
17. Samii A, Dahlen DD, Spence AM, Maronian NC, Kraus EE, Lennon VA. Paraneoplastic movement disorder in a patient with non-Hodgkin's lymphoma and CRMP-5 autoantibody. *Mov Disord* 2003;18:1556–1558.
18. Alexander GE, DeLong MR, Strick PL. Parallel organization of functionally segregated circuits linking basal ganglia and cortex. *Annu Rev Neurosci* 1986;9:357–381.
19. Pujol J, Soriano-Mas C, Alonso P, et al. Mapping structural brain alterations in obsessive-compulsive disorder. *Arch Gen Psychiatry* 2004;61:720–730.
20. Szeszko PR, MacMillan S, McMeniman M, et al. Brain structural abnormalities in psychotropic drug-naive pediatric patients with obsessive-compulsive disorder. *Am J Psychiatry* 2004;161:1049–1056.
21. Kim CH, Koo MS, Cheon KA, Ryu YH, Lee JD, Lee HS. Dopamine transporter density of basal ganglia assessed with [123I]IPT SPET in obsessive-compulsive disorder. *Eur J Nucl Med Mol Imaging* 2003;30:1637–1643.
22. Eng JA, Frosch MP, Choi K, Rebeck GW, Greenberg SM. Clinical manifestations of cerebral amyloid angiopathy-related inflammation. *Ann Neurol* 2004;55:250–256.
23. Fountain NB, Eberhard DA. Primary angitis of the central nervous system associated with cerebral amyloid angiopathy: report of two cases and review of the literature. *Neurology* 1996;46:190–197.
24. Church AJ, Dale RC, Giovannoni G. Anti-basal ganglia antibodies: a possible diagnostic utility in idiopathic movement disorders? *Arch Dis Child* 2004;89:611–614.

Pyramidal Tract Imaging in Multiple-System Atrophy

Nicole Limberg, MBBS,¹ Susanne Jeavons, MBBS, FRACR²
Thomas Robertson, MBBS, FRCPA,³
and Richard Boyle, MBBS, FRACP^{1*}

¹Department of Neurology, Princess Alexandra Hospital, Brisbane, Australia

²Department of Neuroradiology, Princess Alexandra Hospital, Brisbane, Australia

³Department of Neuropathology, Princess Alexandra Hospital, Brisbane, Australia

Abstract: A new radiological finding of T2 FLAIR hyperintensities in the pyramidal tracts is described in a patient clinically thought to have idiopathic Parkinson's disease but histologically proven to have multiple-system atrophy. © 2005 Movement Disorder Society

Key words: pyramidal tract hypertensity; multiple system atrophy

Ramos and colleagues¹ recently reported unusual radiological findings in two patients, one fulfilling the clinical criteria for

*Correspondence to: Dr. Richard Boyle, Department of Neurology, Princess Alexandra Hospital, Ipswich Road, Woolloongabba, Brisbane, 4102, Australia. E-mail: richard_boyle@health.qld.gov.au

Received 1 February 2005; Revised 1 February 2005; Accepted 16 February 2005

Published online 18 July 2005 in Wiley InterScience (www.interscience.wiley.com). DOI: 10.1002/mds.20610

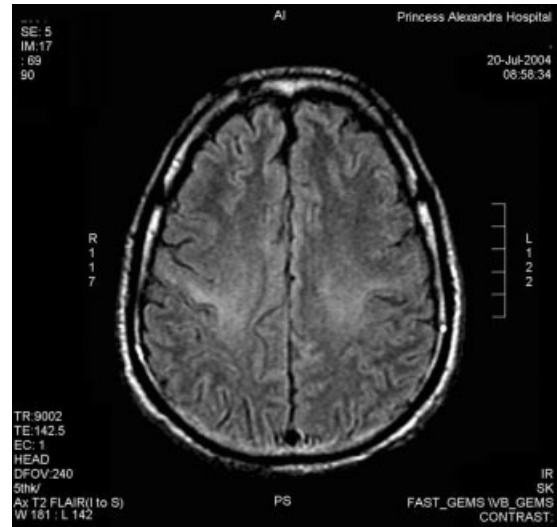


FIG. 1. T2 FLAIR image showing hyperintensities in pyramidal tracts.

probable and one for possible multiple-system atrophy (MSA). We now report identical radiological findings in a case of histologically proven MSA.

Case Report

The patient, a male of Samoan descent, presented at age 36 years with a rigid akinetic syndrome, first affecting his left upper limbs but rapidly becoming generalized. This was initially very sensitive to levodopa. After a couple of years of treatment, dyskinesias became a major problem and over the last year of his life he was virtually bed-bound with alternating periods of profound akinesia and rigidity and severe dyskinesia. He was admitted 1 month prior to his death because of the

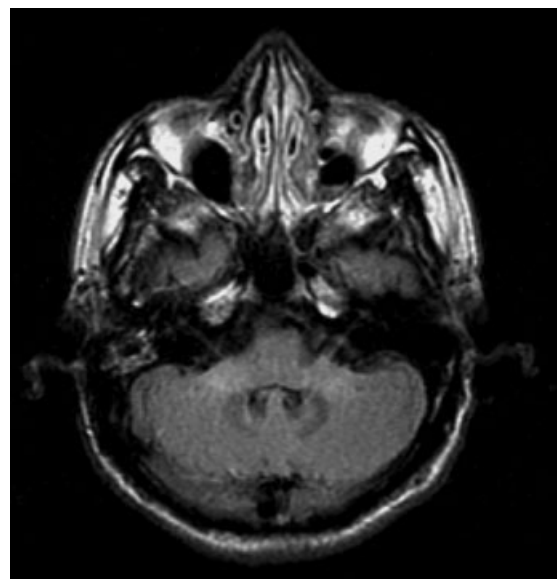


FIG. 2. T2 FLAIR image showing hyperintensities in middle cerebellar peduncles.