SHORT REPORT

Optic nerve and spinal cord manifestations of malignant atrophic papulosis (Degos disease)

F Matsuura, K Makino, T Fukushima, N Matsubara, M Shibuya, T Higuchi, H Hashidate, M Yamada, H Shibuya, M Yamazaki

J Neurol Neurosurg Psychiatry 2006;**77**:260–262. doi: 10.1136/jnnp.2005.066134

A fatal case of malignant atrophic papulosis (Degos disease) with optic nerve and spinal cord involvement is described. Magnetic resonance imaging (MRI) of the optic nerve showed abnormal signal enhancement on fat suppressed T1 weighted images after intravenous meglumine gadopentetate infusion. On T2 weighted sagittal images, a sawtooth pattern was observed over seven vertebral segments of the spinal cord. On necropsy, a severe loss of myelinated nerve fibres in the left optic nerve was seen, with thrombotic obstruction of the central retinal artery. Spongy degeneration was observed in all levels of the spinal cord, with patchy and moth-eaten patterns caused by thromboses and endothelial proliferation in subarachnoid vessels. Findings on MRI were consistent with findings on pathological examination.

Malignant atrophic papulosis, or Degos disease, is a very rare dermatological disorder. When associated with the gastrointestinal tract or central nervous system (CNS), patients with Degos disease have a poor prognosis and a high mortality.^{1–3} There have been few reports documenting magnetic resonance imaging (MRI) findings of CNS involvement in Degos disease.⁴ Generally, the pathology of CNS involvement in a patient with Degos disease includes disseminated occlusive vasculopathy and the resulting diffuse ischaemic changes. There have been no reports where MRI of CNS lesions in patients with this disease were compared with the pathological findings. Moreover, there have been no reports of MRI studies on the optic nerve in patients with Degos disease. Here we present the necropsy results from a patient with Degos disease and compare them with the MRI findings.

CASE REPORT

The patient was a 58 year old Japanese woman. In December of 2002, she visited the Department of Ophthalmology at Niigata City General Hospital with a visual field defect in her left eye, and was diagnosed as having left retrobulbar optic neuritis. Multiple sclerosis was suspected and she received two courses (of three days each) of pulse methylprednisolone therapy. However, the treatment was ineffective and the patient lost the vision of her left eye in February 2003. In mid-June 2003, she developed dysuria and dyschesia, as well as weakness and sensory disturbances in the bilateral lower extremities, followed by a gradual progression of gait disturbances. She was admitted to our department on 9 July 2003.

Physical examination revealed asymptomatic multiple papules (3–6 mm in diameter with a porcelain white centre) on the trunk and proximal extremities. Neurological examination showed loss of vision in the left eye, muscle weakness and exaggerated deep tendon reflexes in the lower

extremities, sensory disturbance below Th10, and vesicorectal dysfunction.

On admission, the patient's blood cell count, biochemistry, serological tests, and coagulation and platelet aggregation activity were all within reference ranges. Antinuclear antibodies and tumour markers were negative. The cerebrospinal fluid lymphocyte count was 2/μl and there was a markedly raised protein level (299 mg/dl). Myelin basic protein and oligoclonal bands were negative.

MRI of the left optic nerve, done in February 2003, showed abnormal signal enhancement on fat suppressed T1 weighted images after intravenous meglumine gadopentetate infusion (post-contrast Is-T1WI) (fig 1A). Brain MRI showed T2 weighted high intensity signals in the right cerebellar hemisphere within the right posterior inferior cerebellar artery territory (fig 1B). T2 weighted MRI of the thoracic cord revealed focal high intensity patterns, with a moth-eaten appearance on axial images (fig 1C), and irregular high intensity lesions between Th5 and Th11 with a sawtooth appearance on sagittal images (fig 1D). However, no gadolinium enhanced lesions were observed on the patient's MRI findings with the exception of those seen in the left optic nerve.

On admission, we thought that the patient had multiple sclerosis, with neuromyelitis optica, because of the transverse myelopathy and the history of retrobulbar optic neuritis-like symptoms. However, while attempting to make a definitive diagnosis we noticed the skin eruptions and carried out a biopsy of the lesions. The biopsy showed thrombosis in the small dermal vessels and intimal hyperplasia. Epidermal atrophy, myxomatous degeneration, and focal necrosis in the dermis were also observed, which appeared to reflect infarctive changes secondary to vessel occlusion. These findings led to the diagnosis of Degos disease, with left optic nerve dysfunction and myelopathy as the CNS manifestations. The patient received antiplatelet and anticoagulant therapy, but developed complete paraplegia by 26 July 2003. On 31 July she developed rebound tenderness and free air on abdominal radiographs, suggesting intestinal perforation. She developed septic shock and died on 29 August, despite vasopressor therapy and respiratory care.

At necropsy, the abdominal organs were clustered together owing to severe peritonitis. CNS examination showed small haemorrhagic lesions in the left basal frontal lobe, primarily in the cortex, which were shown histologically to be fresh haemorrhagic infarcts. Fresh thromboses were also observed in blood vessels in the subarachnoid space. In contrast, old infarcts were scattered in the cerebellum, especially in the right posterior inferior cerebellar artery territory, and blood vessels with organised thromboses were occasionally found in the subarachnoid space.

The patient's optic nerves were found to have degenerated bilaterally, with the degeneration of the left optic nerve being so severe that most myelinated nerve fibres were absent (fig 1E).

Figure 1 (A) Fat suppressed T1 weighted magnetic resonance imaging (MRI) after intravenous meglumine gadopentetate infusion, showing abnormal signal enhancement of the left intraorbital optic nerve (arrow). (B) T2 weighted axial MRI, showing high intensity signals in the right cerebellar hemisphere (arrow). (C) T2 weighted axial MRI, showing irregular focal lesions with high intensity of the spinal cord with a moth-eaten appearance (arrows). (D) T2 weighted sagittal MRI, showing high intensity signals between Th5 and Th11, particularly on the dorsal side, with a sawtooth appearance (arrows). Histopathological changes of the left optic nerve (E, F, G). (E) Klüver-Barrera (KB) staining showing severe degeneration. (F, G) Haematoxylin and eosin (H&E) staining showing complete obstruction by organised thromboses of the central retinal artery (arrow). Histopathology of the spinal cord (H, I). (H) KB staining. Ischaemic lesions of varying sizes are present at all levels of the spinal cord, primarily involving the white matter (arrows). (I) H&E staining. Fresh thromboses and endothelial proliferation are seen in the subarachnoid vessels (arrow).

There were no glial scars or evidence of inflammatory cell infiltration in the optic nerves. The central retinal artery was completely obstructed by organised thromboses (fig 1F, G).

A spongy degeneration with a patchy appearance, probably reflecting ischaemic changes, was observed in all levels of the

spinal cord, primarily involving the white matter in the lateral and posterior columns (fig 1H). Focal areas of myelin and axonal loss were identified in the ventral and dorsal nerve roots. Fresh thromboses and endothelial proliferation were occasionally observed in subarachnoid vessels (fig 1I).

DISCUSSION

The symptoms in this case were characteristic of a neuropathy that began with retrobulbar optic neuritis-like symptoms and progressed to myelopathy six months later. Spatial and temporal variation in the neurological symptoms suggested multiple sclerosis. However, the patient was unresponsive to steroid treatment. In addition, myelin basic protein and oligoclonal bands were absent, and the imaging findings were not consistent with multiple sclerosis. Our patient was diagnosed as having Degos disease on the basis of a skin biopsy, and we believe that her CNS symptoms were a result of this disease. Unfortunately, she was unresponsive to any treatment. Her condition progressed and was fatal within one year and three months after the appearance of skin lesions.

It has been pointed out previously that Degos disease involves optic nerve damage,³ but there have been no detailed case reports about this. In our patient, MRI of the optic nerve, done after the loss of vision in her left eye, showed overall signal enhancement in the left intraorbital optic nerve on post-contrast Is-T1WI (fig 1A). Complete obstruction of the central retinal artery (fig 1F, G) resulted in full length and full thickness degeneration of the intraorbital optic nerve (fig 1E), which may have generated the distinct images seen on post-contrast Is-T1WI, even though the lesion was not active at the time of the MRI study.

Brain MRI showed an old infarct in the right cerebellar hemisphere (fig 1B) at the time of admission, even though the patient had been asymptomatic. Necropsy examination showed a fresh haemorrhagic infarction in the left basal frontal lobe, an old infarct in the right cerebellar hemisphere, and intravascular thrombi in the subarachnoid space. Cerebrovascular accidents such as cerebral infarction, cerebral haemorrhage, and subarachnoid haemorrhage are common types of brain lesion in patients with Degos disease.^{3 5-8}

T2 weighted sagittal MRI of the thoracic spinal cord revealed high intensity lesions with a sawtooth appearance between Th5 and Th11 (fig 1D). T2 weighted axial images of the thoracic spinal cord showed high intensity signals with a moth-eaten appearance in the periphery of the spinal cord (fig 1C). Patchy lesions from the periphery to the centre of the spinal cord, a characteristic of Degos disease,^{5 7} are the likely explanation for these MRI findings. Although the patient's neurological symptoms were consistent with transverse myelopathy below Th10, neither imaging nor pathological findings reflected this pathology. As the lesions were distributed in patchy and moth-eaten patterns on transverse sections (fig 1H), the longitudinal nerve fibres appeared to be

severed by the lesions, resulting in overall symptoms of transverse myelopathy.

Thus the MRI findings in the optic nerve, brain, and spinal cord were consistent with the pathological findings. Secondary occlusion of the lumen resulting from intimal hypertrophy or intraluminal thrombus formation within the small arteries is often seen in Degos disease; however, the aetiology of the disorder is unknown. Such occlusions lead to dysfunction of multiple organs because of ischaemic changes and microbleeding.⁸

A rash is usually the first recognisable symptom in Degos disease, but other symptoms may precede the skin manifestations, posing difficulties in diagnosis. When CNS symptoms are observed, as in this case, MRI of each potential lesion is useful for a differential diagnosis.

Authors' affiliations

F Matsuura, K Makino, T Fukushima, N Matsubara, M Yamazaki, Department of Neurology, Niigata City General Hospital, Niigata, Japan

M Shibuya, Department of Dermatology, Niigata City General Hospital
T Higuchi, Department of Radiology, Niigata City General Hospital

H Hashidate, H Shibuya, Department of Pathology, Niigata City General Hospital

M Yamada, Pathological Neuroscience Branch, Brain Research Institute, Niigata University, Niigata, Japan

Competing interests: none declared

Correspondence to: Dr Kunihiko Makino, Department of Neurology, Niigata City General Hospital, 2-6-1 Shichikuyama, Niigata 950-8739, Japan; makinoncgh@hosp.niigata.niigata.jp

Received 21 February 2005

In revised form 5 May 2005

Accepted 30 June 2005

REFERENCES

- 1 Köhlmeier W. Multiple Hautnekrosen bei Thrombangiitis obliterans. *Arch Dermatol Syphilol* 1941;**181**:783-92.
- 2 Degos R, Delort J, Tricot R. Dermatitis papulosquameuse atrophique. *Bull Soc Fr Dermatol Syphiligr* 1942;**49**:148-50.
- 3 Degos R. Malignant atrophic papulosis. *Br J Dermatol* 1979;**100**:21-35.
- 4 Yoshikawa H, Maruta T, Yokoji H, et al. Degos' disease: radiological and immunological aspects. *Acta Neurol Scand* 1996;**94**:353-6.
- 5 McFarland HR, Wood WG, Drown BV, et al. Papulosis atrophicans maligna (Köhlmeier-Degos disease): a disseminated occlusive vasculopathy. *Ann Neurol* 1978;**3**:388-92.
- 6 Dastur DK, Singhal BS, Shroff HJ. CNS involvement in malignant atrophic papulosis (Köhlmeier-Degos disease): vasculopathy and coagulopathy. *J Neurol Neurosurg Psychiatry* 1981;**44**:156-60.
- 7 Label LS, Tandan R, Albers JW. Myelomalacia and hypoglycorrhachia in malignant atrophic papulosis. *Neurology* 1983;**33**:936-9.
- 8 Subbiah P, Wijedicks E, Muentner M, et al. Skin lesion with a fatal neurologic outcome (Degos' disease). *Neurology* 1996;**46**:636-40.