autonomic failure. On 21 July, she had a low grade fever, and was transferred to Kishiwada Hospital. Her temperature was 37.5°C and she had a normal serum creatine kinase concentration. Five days later, she developed a pyrexia of 42°C with high serum creatine kinase (1500 IU/l). Intravenous dantrolene therapy produced no response and she died of disseminated intravascular coagulation two days later.

In these six episodes of an NMS-like condition, one was caused by decreased antiparkinson medication, one by an antidepressant drug, two by complications, one by hyperthermia from environmental origin, and one by hyperthermia with L-threo-DOPS. Five episodes occurred in the hot season. In Japan, we had hot days in October 1995. Two cases had multiple episodes.

A putative mechanism of pyrexia in an NMS-like condition is dopamine depletion in the extrapyramidal system causing severe rigidity, resulting in high fever and creatine kinase leakage from the muscle to the blood.¹ With antipsychotic drugs, the stronger the affinity to the D₂ receptor, the more often NMS occurs. On the other hand, some agents which do not influence the dopaminergic system also produce an NMS-like condition. An abnormal mechanism in the monoaminergic system is thought to alter the function of the thermal centre of the hypothalamus.³ The setting of body temperature becomes higher by activation of the noradrenergic system, and lower by the dopaminergic or serotonergic systems in the thermal centre.⁴ An imbalance of these systems via medication or other causes may trigger an NMS-like condition. In patient 1, an antidepressant, which influences serotonergic and noradrenergic activities, caused the second episode.

It is characteristic that five episodes were caused by somatic stresses such as infection or hyperthermia of various origins. This condition occurred during the hot season in patients with or without antiparkinson drugs. In a patient with vascular parkinsonism not receiving antiparkinsonian drugs, an NMS-like condition after typical hyperthermia⁵ was reported to be immediately alleviated by intravenous levodopa therapy. This suggests that activation of the dopaminergic system was important in the NMS-like condition even when dopamine depletion was not its cause. In patient 3, treatment of pyrexia with L-threo-DOPS, a precursor of noradrenaline, caused a further increase in body temperature and development of an NMS-like condition. This substance is effective for akinesia in Parkinson's disease and dysautonomia, and is reported to provoke an NMS-like condition by causing monoaminergic imbalance in the CNS.² A change in hypothalamic noradrenergic activity caused by L-threo-DOPS seems to precipitate an NMS-like condition.

Besides withdrawal of dopaminergic agents, administration of agents which alter monoaminergic neuron activities, stresses to the body, and heat increase from various causes tend to be triggers of an NMS-like condition. Patients with multiple system atrophy, which usually affects thermal regulation, are particularly at risk of developing the condition.

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Non-Hodgkin's lymphoma as a new cause of non-thrombotic superior sagittal sinus occlusion

The syndrome of non-thrombotic superior sagittal sinus occlusion is an uncommon complication of local neoplastic disease which presents clinically as chronic intracranial hypertension without focal signs.¹ A few cases have been reported as complications of midline tumours involving the occipital region such as Ewing's sarcoma, plasmacytoma, neuroblastoma, and disseminated carcinoma of the breast.¹⁻⁴

To our knowledge, this is the first case report in which this complication appeared secondary to a non-Hodgkin's lymphoma.

A 36 year old man presented with a subcutaneous mass in the occipital region of the scalp which had increased in size during the past year. His medical history was unremarkable.

The tumour was painless, hard, and fixed to underlying tissue. Neurological examination was normal, except for bilateral papilloedema. Visual acuity was normal. Cranial MRI disclosed an occipital subcutaneous tumour 10×8×3 cm in diameter. Surrounding bone was thickened, and part of the mass extended extradurally, causing displacement of the venous sinuses. Normal signal void in the superior sagittal sinus in T1 weighted images was replaced by a hyperintense signal (figure, A). Complete coagulation studies were normal. The tumour biopsy disclosed a non-Hodgkin's lymphoma and extension study showed a metastatic lesion in the right femur, confirmed by biopsy.

The patient was treated with local radiotherapy and chemotherapy (six sessions CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone)).

The subcutaneous mass had disappeared two months later. At one year follow up the patient did not complain of neurological or ophthalmological symptoms, and recent MRI and magnetic resonance angiography showed the absence of occipital tumour and superior sagittal sinus occlusion (figure, B).

(A) Sagittal cerebral T1 weighted (TR 441, TE 15) MRI (1 tesla) showing an occipital subcutaneous tumour infiltrating bone and extending extradurally causing occlusion of superior sagittal sinus (arrow); note a retention cyst in sphenoid sinus. (B) Sagittal cerebral T1 weighted (TR 600, TE 15) MRI (1.5 tesla) one year after local radiotherapy and chemotherapy: the mass has disappeared and superior sagittal sinus has returned to normal.

The syndrome of non-thrombotic occlusion of the superior sagittal sinus, first described by Plant *et al.*¹ consists of compressive occlusion of dural venous sinuses secondary to midline tumours in the occipital region of the skull. The clinically distinctive feature is the lack of focal signs in the course of chronic intracranial hypertension.

The most common thrombotic occlusion of the superior sagittal sinus is an acute disorder, usually with disturbance of consciousness and focal signs, including seizures and paresis.⁵ Reported cases of occlusion of the superior sagittal sinus associated with neoplastic disease have usually been examples of thrombotic complications secondary to hypercoagulability states.

The syndrome of non-thrombotic occlusion of the superior sagittal sinus occurs when a tumour produces an extradural tissue mass beneath the inner table of the skull, and it is difficult to differentiate between compressive occlusion of the superior sagittal sinus or tumour invasion of the dura and sinus. Cerebral angiography may help in such differentiation, but unfortunately, this examination was not performed in our patient.

This complication has not been described in tumours that commonly give rise to bone metastases such as carcinoma of the bronchus, which develop expanding lytic lesions in the skull with minimal soft tissue involvement. Previous cases of non-thrombotic occlusion of the superior sagittal sinus have been reported secondary to solitary plasmacytoma, Ewing's sarcoma, disseminated carcinoma of the breast, and neuroblastoma,¹⁻⁴ which, as in our case, all present significant involvement of soft tissue.

It is also worthy of mention that the site of first clinical manifestation in our patient, the skull, is suggestive of metastases rather than that of a primary intracranial neoplasm as confirmed on further scanning. Moreover, the excellent response to standard chemotherapy suggests a systemic vascularisation of this tumour.

The site of compression of the superior sagittal sinus in nearly all the reported cases, including the present patient, is the terminal portion of the superior sagittal sinus and the torcular herophili.¹ The chronic course of the sinus compression allows the formation of good collateral venous return, making this process clinically silent.

In conclusion, we report a new cause of non-thrombotic occlusion of the superior

sagittal sinus, and emphasise that this syndrome should be considered in cases of mid-line occipital neoplasms presenting with raised intracranial pressure without focal signs.

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