occurred every three or four seconds associated with eye blinking. All tendon reflexes were pathologically brisk with bilateral extensor plantar responses.

EEG showed stereotyped repetitive slow wave disturbances which were linked in time to the myoclonic jerks, characteristic of SSPE. CT scan showed areas of poorly defined low attenuation in white matter of both frontal lobes, the left temporal horn and near the trigones. Cerebrospinal fluid (CSF) analysis revealed protein 690 mg/l; normal IgG concentration and oligoclonal bands were detected on polyacrylamide gel electrophoresis. Measles antibody titres (complement fixation test) were raised in the CSF (1:64) and serum (1:1024).

He had been treated with clonazepam, phenytoin and dexamethasone, and subsequently with isoprinosine, but his condition has continued to deteriorate. He is now (May 1982) unresponsive. The myoclonic jerks have become less marked.

This patient developed classical clinical features of SSPE with progression from the stage of insidious personality change to that of visual failure, pyramidal rigidity with myoclonic jerking of the limbs, proceeding to a state of mutism, incontinence and complete paralysis. There was a history of measles infection in childhood. The onset of SSPE at the age of 17 years is later than usual although well recognised. The investigations were typical of those found in SSPE. It seems reasonable to assume that the earlier symptom of transient homonymous hemianopia was part of the same illness, especially in view of the fact that his initial visual symptoms in November 1981 were very similar to those which had occurred nearly two years previously. As far as we are aware, the presentation of SSPE with transient homonymous hemianopia has not been described previously, and the delay of nearly two years between the onset of the hemianopia and the subsequent development of classical SSPE symptoms is remarkable. The disease is usually fatal within one or two years of clinical onset, although some patients have survived for much longer periods. The total duration of this patient’s illness is not yet known. In this case the visual loss which occurred subsequently was probably due to cortical blindness, which is consistent with observations that neuronal loss in SSPE is very marked in the occipital lobes. If this pathological process had indeed occurred at the onset of the illness in January 1980, it remains unclear why it should have been arrested for a period as long as two years.

PGE KENNEDY

References


Sagittal sinus thrombosis and occult malignancy

SIR: We describe a patient with sinus thrombosis diagnosed in life, the cause at necropsy being intravascular metastases from a carcinoma of the gall bladder. In contrast to previously reported cases, there was no manifestation of malignancy in life.

A lady, aged 58 years, had been treated in 1972 with radio-iodine for thyrotoxicosis. The proptosis present at the time did not resolve. In 1980 she presented with a two month history of headache, temporal pain and malaise. She was admitted to her District General Hospital where, over a week, she became drowsy, confused and complained of double vision. An isotope brain scan suggested a subdural collection and she was transferred to the regional neurology unit on dexamethasone therapy. At transfer she appeared pale and unwell. The blood pressure was 210/90, there was a soft systolic ejection murmur at the left sternal edge without radiation or signs of endocarditis. She was clinically euthyroid, had bilateral propotosis, sixth nerve palsy and papilloedema. Hearing was reduced by a bilateral conductive defect. There was no pyramidal signs and there was no meningism. After four days, hypoaesthesia to light touch and pin prick was noted on the lower lip. The proptosis became worse, periorbital oedema and chemosis appeared bilaterally. She developed a flaccid left hemiparesis, bilateral extensor plantar responses and continued to deteriorate, bleating from venepuncture sites; she died on the tenth day of admission.

Serious blood cultures were sterile, serology normal, ESR raised at 70 mm/h and temporal artery biopsy normal. Left ventricular hypertrophy was evident on the ECG; radiographs showed a small left pleural effusion and generalised degenerative changes of the spine on skeletal survey. Cranial computed tomography demonstrated an old frontal lobe infarction and no evidence of a subdural collection. The cerebrospinal fluid was under increased pressure at 280 mm and was clear. The cell count, electrophoresis and cytocentrifuges were normal and the protein 48 g/l. Cavernous sinus thrombosis and possibly sagittal sinus thrombosis was suspected but angiography could not be performed because of disseminated intravascular coagulation.

At necropsy the dura was inflamed and densely adherent to the skull. The sagittal sinus was thrombosed and the carotid arteries inflamed in the cavernous sinus. Anaplastic cell nests were present in the artery walls, in the organising thrombus in the sagittal sinus and in the sphenoidal sinuses. Tumour was also present in the pontine veins. The gall bladder wall was thickened and haemorrhagic; the capillaries were packed with anaplastic tumour. Similar changes were seen in the liver, pancreas and para-aortic lymph nodes. The cells were of biliary origin and the primary site thought to be gall bladder.

Sagittal sinus thrombosis may be associated with infection, dehydration, head injury, leukaemia, malignancy and obstetric problems such as abruptio placenta. As in this case headache, papilloedema, ophthalmoplegia and paraparesis suggest the diagnosis. Additional features include nausea, vomiting, convulsions, hemiparesis, paraparesis, visual field defects and nuchal rigidity. The cerebrospinal fluid is under increased pressure and contains excess protein and a raised white cell count. The peripheral white cell count is often raised. CT scanning may be
unhelpful as in this case, unless venous infarction of the cerebrum or cerebellum has occurred. The diagnosis may be made by carotid angiography or dynamic isotope scanning. The EEG is frequently abnormal showing a focal or widespread abnormality but is not diagnostic.

Sagittal sinus thrombosis has been described previously with malignant disease for example with nephroblastoma, Ewings tumour, carcinoma of the breast and leukaemia. All diagnoses were established before the onset of neurological symptoms, the necropsy findings being either non-metastatic sinus thrombosis, extravascular deposits or, less commonly, tumour present within the thrombus. Some patients in these series had additional contributory factors such as bacteraemia, recent radio-therapy and chemotherapy. Carcinoma of the gall bladder occurs with an incidence of 2.5:100,000 of the population. It is more common in females (1.4:1), the maximal incidence being the seventh decade for females and the sixth for males. The tumour may simulate chronic cholecystitis with which it is associated or may cause many and varied symptoms, even presenting an acute abdomen with perforation or intestinal obstruction. Because of this the diagnosis is frequently made late and the prognosis is therefore poor. The majority of these patients are discovered at surgery and are adenocarcinoma in 80%, anaplastic in 10% and the remainder squamous cell or benign. Disseminated intravascular coagulation is seen in many clinical situations, and has been described in malignant disease particularly in the terminal phases and when associated with infection, chemotherapy and radiotherapy.

It is unusual for sagittal sinus thrombosis to occur in association with an undiscovered primary tumour. Disseminated intravascular coagulation prevents full investigation during life, but it is unlikely that the cause would have been found. It is important to consider disseminated malignancy as a possible cause of sagittal sinus thrombosis when no primary tumour is obvious.

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