Ospocclus as a presenting symptom in thymic carcinoma

Ospocclus is a rare paraneoplastic syndrome accompanying some malignant tumours (carcinoma of the lung, uterus, breast, and neuroblastoma in children). We report a patient with ospocclus combined with ataxia preceding the diagnosis of thymic carcinoma.

At presentation the patient complained of dizziness and difficulties with walking before admission to hospital. This was followed by episodes of nocturnal sweating and diffuse abdominal pain. Medical, neurological and family history was unremarkable. On admission examination showed the patient to be in a good general condition. The neurological examination revealed normal consciousness; speech and oculo-motor function were normal. The most relevant finding was the patient’s eye movements; there were rapid, chaotic and conjugate saccades of the eyes in all directions, considerably exacerbated by atropine. The red discs were normal in size, reactive to light and accommodation and the fundoscopy showed normal optic discs. The examination of the rest of the cranial nerves was normal. Myoclonic seizures were not seen. Superficial and deep reflex testing did not reveal any abnormalities.

Motor and sensory examinations were normal except for his gait which showed mild trunk ataxia. Finger-nose test, heel-to-toe and rapid alternating manoeuvres were within normal limits. The values of the laboratory tests including blood glucose, electrolytes, urea, uric acid, bilirubin, creatinine, serum albumin and globulin, calcium, phosphate, SGOT, alkaline phosphatase, ESR, haemoglobin and leukocytes were within normal limits. Beta-human chorionic gonadotropin in the blood, alpha-feto-protein and 5-hydroxy-indole acetic acid in the urine were also within normal limits. The level of blood immunoglobulins was normal. Serological tests for syphils were negative. The bone X-rays revealed two mediastinal masses. Computerised tomography (CT) of the chest showed two masses in the right anterior mediastinal space attached to the pericardium. Electroencephalogram, ECG, skull radiograph, thyroid and liver scans and two CT scans of the brain were within normal limits. Lumbar puncture showed normal CSF pressure. A CSF analysis for total protein, glucose, chloride and cell count was normal. The auditory brain stem evoked potentials. Bone biopsy and bone marrow did not reveal any malignant cells. The Tension test was negative.

One month after admission the patient had a right anterior mediastinotomy for biopsy of the masses. The histology revealed mixed thymic carcinoma (lymphoepitheliomata type, poorly differentiated squamous cell carcinoma) with pleural metastases. The patient was treated with a combination chemotherapy regime which is used in the treatment of malignant thymoma. Three weeks later there was a further deterioration in the patient’s general condition. Chest radiograph showed no change and CT scan of the brain was within normal limits. Chemotherapy treatment was continued with no subjective or objective change.

Following the failure of chemotherapy, the patient was treated with radiotherapy but with no improvement. The patient was discharged, but readmitted again two months after discharge for marked general and neurological deterioration, with severe ataxia, bilateral dysmetria, ospocclus and chest pains. A trial treatment with clonazepam 2 mg/day orally for seven weeks had no effect on the ospocclus or ataxia, and this treatment was discontinued. His general condition deteriorated further and he died nine months after the onset of the symptoms of the disease. Permission for necropsy was refused.

Ospocclus may occur in encephalitis and as a remote effect of systemic malignancy, such as neuroblastoma in children, carcinoma of the lung, breast, uterus and thyroid.1 This phenomenon has been recently reported in cases of demyelinating disease, brain glioblastoma, thalamic haemorrhage, hydrocephalus, Friedreich’s ataxia, intoxication with lithium and chemotherapy-induced hypoglycaemic states.2 In our patient, the ospocclus was accompanied by truncal and limb ataxia. These symptoms were the first manifestation of carcinoma of the thymus. The association of ataxia with thymic carcinoma has not been previously reported.

The site of the lesion that causes ospocclus remains unclear. The association of myoclonus and a cerebellar syndrome in a few cases, indicated that the lesion is in the inferior vermis of the cerebellum in the connection between the inferior olives, red nuclei and the dentate nuclei.2 However, there is no evidence that either a lesion in the dentate nuclei or in the red nuclei produces ospocclus by itself.

In a few cases of osppocclus and systemic carcinoma, the necropsy studies reported only inconsistent pathological findings of poor or localising value, such as a mild loss of Purkinje cells, a peridental gloss and slight loss of myelin in the cerebellum.3 In other patients, perivascular infiltrates and gliosis in the pons, midbrain and hypothalamus were described. In a few patients necropsy studies had no demonstrable pathology in the brain or cerebellum.

The pathophysiological mechanisms of the lesions that cause ospocclus remain unknown. The various viral infections of the CNS (as in cases of ospocclus in patients with encephalitis) or immunological dysfunction (as in demyelinating disease or in malignant disease) may be involved in the process. In support of the immunological theory are the elevated levels of CSF-IgG in some cases,4 the effective response to corticosteroid therapy in others5 and the presence of specific antibodies to the Purkinje cells that have been found in two patients with systemic malignancy.6 In ospocclus caused by a paraneoplastic syndrome, neurological signs fail to improve after treatment. In certain cases, clonazepam and thiamine were effective.6 In our case, treatment with corticosteroids and clonazepam did not change or improve the evolution of the neurological signs.

Standard neurodiagnostic tests in Sydenham’s chorea

The recent resurgence of acute rheumatic fever in the United States has served to focus attention on the procedures for diagnosing Sydenham’s chorea (SC). No single test is available for this purpose and it is widely recognised that a rheumatic etiology often cannot be established with certainty at the time of presentation of chorea.2 In the era when acute rheumatic fever in the developed world was sufficiently common for large numbers of patients with SC to be collected, the standard neurodiagnostic tests were limited to electroencephalography (EEG) and analysis of cerebrospinal fluid (CSF) for total protein and white cell count.2 Futhermore, few alternative causes of the chorea were sought.

We report here a series of three contemporary neurodiagnostic tests in a prospective study of SC.

The subject group comprised five consecutive cases seen by the authors at Kalamfong Hospital over a period of three years (January 1985 to December 1987). Kalamfong is a general hospital on the outskirts of Pretoria serving the black community. All the patients were males, their ages ranging from seven to 16 years. Serologic evidence of recent streptococcal infection (antistrep-tolysin-O titre > 200 Todt units, or antistreptolysin-O titre > 1000) was sought. We report here the results of a series of contemporary neurodiagnostic tests in a prospective study of SC.

Each patient fulfilled the following criteria, anamnestic details being provided or corroborated, by a parent or close relative. 1 Involuntary movements typical of chorea developing insidiously in the setting of previously good health. 2 Alert mental state. 3 Absence within the family history of a progressively disabling disorder. 4 Negative history for drug ingestion preceding, or concurrent with, the onset of chorea. 5 Normal test values for thyroid, renal and hepatic function. 6 Normal full blood count and red cell morphology. 7 Normal plasma glucose and serum sodium, calcium and phosphate. 8 Absence of Kayser-Fleischer rings, normal serum copper and ceruloplasmin. 9 Negative tests for rheumatoid factor, anti-nuclear antibodies, neurophilys (specific and non-specific). 10 Negative pregnancy tests (where indicated).

Cerebrospinal fluid analysis, contrast enhanced computed tomography (CT) brain scan and upper (median nerve) and lower limb (tibial nerve) cortical somatosensory evoked potentials (SEP) were normal in all

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