chronic hemidystonia following acute dystonic reaction to thiothylperazine

Thiothylperazine is a phenothiazine neuroleptic drug that blocks postsynaptic dopamine D2 receptors and is extensively used for the treatment of vertiginous syndromes. Its extrapyramidal side effects include acute and tardive dystonic reactions and a Parkinsonian syndrome. The acute reactions are usually self-limiting and brief and need treatment with anticholinergic therapy. We report an unusual case of a chronic hemidystonia which began immediately after treatment with thiothylperazine.

A 47 year old man, with no previous personal or family history of neurological illness, was evaluated in December 1988 in our unit because of involuntary movements and posturing of his right limbs. When he was aged 40 he developed a vertiginous syndrome (retrospectively diagnosed as “vestibular neuritis”) and was treated with thiothylperazine 6.5 mg three times daily. When he was aged 44 he developed dystonic movements and postures of the right limbs, neck and trunk that persisted until the day of his admission. Apart from his movement disorder, the general and neurological examination were normal. A blood cell count, biochemistry, urinalysis and copper studies in plasma and urine, were all normal, and serological tests for syphilis were negative. EEG, cranial CT and MRI showed no abnormalities. The dystonic movements improved moderately with biperiden 8 mg/day, although he has a persistent action dystonia of his right limbs, which is more noticeable during walking.

Dystonia has been classified according to distribution as focal, multifocal, segmental, or generalised, and when its distribution involves the ipsilateral arm, leg, and face, the term “hemidystonia” is applied. The presentation of a dystonic syndrome of hemidystonia has an important aetiological significance because in most cases there is CT evidence of contralateral basal ganglia damage. The lack of evidence of a structural lesion in the basal ganglia in our patient on CT and MRI, together with the onset of this clinical picture immediately after thiothylperazine exposure, implies that hemidystonia may be caused by a dystonic engram (in hemidystonia) in a subject possibly already predisposed to develop idiopathic dystonia) by phenothiazine.

Acute dystonic reactions after exposure to phenothiazines or other neuroleptic agents usually disappear after withdrawal of the relevant drug and/or anticholinergic therapy. Nevertheless, some cases have been reported in the literature of prolonged or chronic dystonia. The series of tardive dystonia by Burke et al includes 42 patients with dystonia after exposure to antipsychotic drugs. Dystonia developed after a mean interval of exposure of 3-7 years.

In none of these cases nor in recent series of tardive dystonia, the distribution ever hemidystonic. Our case therefore appears to be the first report of hemidystonia occurring as a direct or indirect side effect of a neuroleptic drug.

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Bilateral metastases in the cerebellopontine angle

Only 0.2% of the pontocerebellar angle (PCA) tumours are metastases. To our knowledge we report the first case of bilateral metastasis.

In 1974, a 64 year old woman presented with a malignant melanoma of the left leg which was treated by excision and chemotherapy. An inguinal lymph node metastasis was found in 1976 followed by a subcutaneous metastasis on the right deltoid region six years later were treated surgically. From June to July 1987 the patient experienced a right-sided hearing loss, an unsteady gait with vertigo, a swallowing impairment and dysphonia. The brain CT scan and the CSF analysis were unremarkable. The patient was admitted to our hospital on 20 July because of a right facial palsy and left hypacosia.

On the right side, there was involvement of the VII–Vth cranial nerves. On the left, Vth, VIth and VIIIth cranial nerves, were involved slightly. The patient appeared ill on skin examination there was no evidence of malignant melanoma.

The CT scan (fig a) showed two hyperdense lesions which on MRI (fig b) were isointense during T1 and hyperintense during T2. The first image, which was round, was in the right PCA and disrupted the brain stem and the cerebellar without invading them; the second, which was oval and smaller in size, was in the left PCA, without mass effect.

On 10 August 1987 a soft, haemorrhagic, yellowish, self contained (2 cm x 3 cm) tumour was found in the right PCA, disrupting the last cranial nerve, forcing back the brain stem on the median line, and spreading onto the jugular foramen. An identical tumour on the left (1.5 cm diameter) did not press on the brain stem but spread along the last cranial nerves without disrupting them.

The histopathological examination showed that they were metastases of an undifferentiated cancer; cells of epithelial type were associated with long cells which looked like sarcoma. On electron microscopy, there was no melanosome, but small stick inclusions in the ergastoplasmic cisterns (75% of cells), of variable orientation, often crossed and made from straight microtubules, suggesting a melanoma origin.

The patient died in October 1987. A necropsy examination could not be performed.

Cornelii reported the first case of a metastasis developed in the PCA; the primary lesion was an oropharynx epithelioma. Since then, most of the unusual published cases are isolated ones, originating from histologically differing types such as: lung, breast, oropharynx carcinoma, carcinoma of probably colon origin, histiocytic malignant lymphoma, malignant histiocytosis, Smorhous xanthoma. No case of melanoma was reported.

In our case, there were several inclusions of straight microtubules in the ergastoplasmic cisterns on histopathological examination, similar to those observed in 6% of cases of...
Myasthenia gravis aggravated by pyrantel pamoate

Pyrantel pamoate is an antihelmintic agent used worldwide.1 We report worsening of myasthenia gravis by pyrantel pamoate in one patient.

In October 1989 a 72 year old diabetic man noticed mild bilateral palpebral ptosis when waking from sleep. He was blind in the right eye from diabetic retinopathy. In mid November of the same year, he complained of diarrhoea. A stool specimen was positive for Ascaris lumbricoides and a single 1000 mg dose of pyrantel pamoate was taken orally with breakfast on 26 November. Several hours later he became fatigued when chopping and also when walking. The following week he could not chew meat and bread and he noticed dysphonia. Neurological examination on 4 December showed bilateral palpebral ptosis with fatigability, limitation of abduction of both eyes and limitation of adduction of the right globe, hypophoria, weakness of neck extension and weakness of abduction of both arms.

Administration of 2 mg of edrophonium chloride reversed the ophthalmoplegia. Electrophysiological study revealed a normal area of compound muscle action potential in the right abductor digitii quinti muscle both at rest and after 15 seconds of maximal voluntary effort. Supramaximal repetitive stimulation of the right ulnar nerve at 3 Hz and 30 Hz were within normal limits.2

Stimulation single fibre electromyography3 of the right externus digitorum communis muscle showed a mean jitter of 40 µs (upper normal limit 25 µs) in 50% of insertions showing increased jitter and no blockings were found. Haematocrital and biochemical tests were normal. Antinuclear antibodies were positive at a 1/340 titre with an homogenate positive. Antiacetylcholine receptor antibodies were not determined. Antismooth muscle and antimitochondrial antibodies were positive at a 1/80 titre. CT of the thorax was normal. Antiacetylcholine receptor antibodies were not determined.

He was treated with 180 mg day of pyridostigmine bromide without alleviation of his symptoms. There was no improvement in spite of increasing the daily dose of pyridostigmine and muscarinic symptoms appeared. Prednisone ingestion was started. Two weeks later he could tolerate solid food. Dysphonia, weakness of neck extension and weakness of the extremitities disappeared, but ophthalmoplegia persisted. From February 1990 he was treated with 70 mg every other day, without neuromuscular symptoms apart from ptosis and bilateral ophthalmoplegia.

We feel that this patient, ingestion of pyrantel pamoate aggravated a previously existing myasthenia gravis. Pyrantel exerts its antihelmintic action by blocking the worm's neuromuscular transmission, producing a depolarising-type neuromuscular block.11 In rabbits, parenteral administration leads to paralysis and death,12 but toxic neuromuscular effects in humans have not been reported to date to our knowledge.

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Myxopapillary ependymomas arising from nerve roots of the spinal cord

Conventional pathogenesis suggests that myxopapillary ependymomas arise from ependymal cells lying adjacent to fibrous tissue. The large majority of myxopapillary ependymomas arise from the filum terminale where this arrangement applies. I report two cases of myxopapillary ependymomas arising directly from ependymal cells lying against the nerve roots.

Ependymomas form 2–6% of all gliomas.1 Myxopapillary ependymomas belong to a distinctive subgroup of ependymoma. They are virtually restricted to the cauda equina and are thought to originate from the nerve roots and conus medullaris. Myxopapillary ependymomas form 16% (953 to (21%) 8/27 of intraspinal tumours (six and two respectively). The myxopapillary ependymoma histology has led to the belief that the pathogenesis of myxopapillary ependymoma is related to ependymal cells lying against fibrous tissue mainly the filum terminale.2 However, these cases may not be as rare, as in subcutaneous sacrococcygeal tissues,11 the lateral ventricle,12 and the cervico-thoracic cord.13 The pathogenesis for myxopapillary ependymomas arising in such a location is less clear.

The first case was a 48 year old male with a one month history of low back ache and left leg sciatica with dragging of his left foot. There was weakness of the left ankle dorsiflexion,14 atrophy of the left foot, and loss of vibration in the knee on the same side. Myelogram showed complete block at spinal level lumbar 2. The second case was a 44 year old male who had experienced low back pain for three years. There was no history of sensory loss or motor weakness. Myelogram revealed a block at spinal level thoracic 12.

Macroscopically these were large encapsulated tumours. The myxopapillary ependymoma in case 1 arose from the first sacral root and in case 2 it arose from the second lumbar root. Treatment for both cases was excision of tumour and resection of the nerve root.

The presentation of these two tumours is similar in many ways to myxopapillary tumours arising from the filum terminale or the conus medullaris, namely low back pain with myelographic block.15 The orthodox pathogenesis for myxopapillary ependymoma is from ependymal cells lying against fibrous tissue for example the filum terminale. Other proposed theories include myxopapillary ependymoma arising from ependymal rests in extra dural locations such as the sacrococcygeal region.16 It has been observed1 that ependymal rests occur in normal children, and occur where myxopapillary ependymomas arise in the same manner as the Arnold Chiari malformation, namely the dermis–subcutaneous junction. It may be that myxopapillary ependymomas arise in abnormal sites as a result of heterotopia, but heterotopia of tissue of the central nervous system in sites other than nasal is open to doubt.13 Others13 have suggested that the presence of ependymal cells against fibrous tissue is not essential for the formation of myxopapillary ependymomas and the possible change seen in myxopapillary ependymomas. They suggest that this myxoid change is the result of anoxia due to vascular changes. A Gilmour et al.16 have suggested that the primary cause of myxopapillary ependymoma or the changes seen within them,
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