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Movement disorders in the elderly

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Movement disorders are common in the elderly, and in particular, Parkinson's disease (PD) and essential tremor (ET) may be a source of considerable concern and morbidity.

Essential tremor (ET)

ET is a common condition and is typically of concern to the patient for two major reasons, one relating to the motor disability and embarrassment, and the other out of concern that they may have PD.

With some exceptions, it is generally straightforward to distinguish the two, largely on the basis that patients with ET typically do not have any complaints relevant to bradykinesia, and are able to attribute the motor difficulties entirely to the underlying tremor. Not uncommonly, patients with ET have a very long history incompatible with the reasonable level of motor function that they possess were their illness due to PD.

On examination, distinguishing features to separate ET from PD include normal spontaneous movements, and normal facial appearance.

In patients with ET, there may be some apparent increase in tone, especially when asking the patient to reinforce by moving the contralateral limb. Bradykinesia may be difficult to assess because of the prominent tremor. However, the distribution of tremor and quality of the tremor are typically pathognomonic – in ET the head is sometimes involved (largely, a no-no tremor), and as is widely recognised tremor tends to be symmetrical, and will typically be present largely with posture-holding (arms outstretched in front of the patient), and brought out by action. Rarely, patients with ET will have a degree of rest tremor, but this is uncommon. Note that, although the tremor is certainly bilateral in the vast majority of cases, the degree of symmetry varies. Standard manoeuvres to bring out tremor would include posture holding, asking the patient to draw a spiral or to write, and asking the patient to pour water from one cup into another. Patients with PD certainly may have action tremor and tremor with posture holding, but this is typically markedly unilateral and is associated with rest tremor.

Treatment may range from reassurance and medication to neurosurgery. In general, consensus is that beta-blockers are not particularly effective and certainly have significant side-effects, particularly in the elderly. The drug of choice is likely to be primidone (Mysoline). Significant drowsiness is virtually universal and the vast majority of patients should typically begin with a half or quarter 25 mg tablet at night and gradually taper the dose upwards depending on tolerability. Some patients may require up to 750 - 1 000 mg daily. Alternatively, topiramate may be used, noting common side-effects of neuropsychiatric disorders, and occasionally, word finding difficulty. In some patients, in whom success is not achieved with either of these two medications, bilateral deep brain stimulation may be advisable.

Parkinson's disease

PD is a common disorder in the elderly, with a prevalence of 2 - 3% in the > 65 year-old population, and an incidence which is exponential, particularly in males. Age is the largest known risk factor for the development of PD, and for dementia in PD.

Recent developments in the pathophysiology of PD include the finding that PD is likely to begin in the lower medulla and olfactory bulb and gradually progress up the brainstem, presenting with the typical features of PD (tremor, rigidity, akinesia and postural instability – TRAP) only when reaching the midbrain.[1] Similarly, it appears highly likely that the illness spreads directly from one cell to another.[2] As with many other neurodegenerative disorders, PD, like Alzheimer's disease, is associated with inclusion bodies, and these represent the incapacity of the cell to deal with an increasing protein load, which initially may be walled off, but subsequently overcome the defence mechanisms of cell, resulting in cell injury and death.

PD, as with dementia with Lewy bodies (DOB), is a synucleinopathy, so termed since both disorders are associated with inclusion bodies that stain positively for the protein synuclein, a finding that followed on the identification of synuclein mutations in rare autosomal dominant forms of PD. The 2 conditions are largely distinguished by their target within the central nervous system, PD appearing to spread gradually upwards, whereas DOB involves the cortex from the beginning. However, neuropsychiatric and cognitive impairment is very common in PD, with rates of 40% at 4 years of the illness in a community-based study.[3]

It is unfortunate that there have not been any significant therapeutic advances in the treatment of PD for a number of years. The neurological literature probably tends to overemphasise the problems of dyskinesias and motor fluctuations (on and off). These tend to be less of a problem in the elderly, and certainly, community-based studies suggest that they may frequently be relatively mild and can be improved by mild dose reduction.[4] Certainly, in the majority of elderly patients presenting with PD, the drug of first choice would be levodopa. Anticholinergics are widely recognised to have a range of side-effects that are undesirable, particularly cognitive impairment and constipation, and although they certainly have a place in the treatment of PD, perhaps particularly with tremor, they should be used with caution. Although dopamine agonists have been promoted as first-line treatment, this is probably not advisable in the usual elderly patient, since they are associated with a poor therapeutic profile, notably drowsiness and a greater degree of orthostatic hypotension, compared with levodopa. In addition, the practitioner needs to be aware that dopamine agonists may all be associated with impulse control disorders (ICDs), a phenomenon not limited to the treatment of PD, but also seen with use of dopamine agonists for treatment of restless leg syndrome.[5,6] Although rare,

this side-effect may be highly significant and it is probably appropriate to warn the patient formally of the potential risk of these agents. Fortunately, ICDs resolve on cessation of the drug.

The treatment of tremor may be a particularly frustrating area in PD, particularly since the patient often presents only with this complaint. Unfortunately, most standard treatments are relatively poor at improving tremor, and it is sensible to let the patient know this when treatment is initiated to avoid subsequent disappointment. If tremor is significant and causing impairment of quality of life, three considerations include: the use of anticholinergics, which may be slightly more effective than the usual treatment of levodopa or dopamine agonist; the use of the atypical antipsychotic clozapine, which is often highly effective, but requires monitoring of white cell counts for 18 weeks following initiation of treatment; and neurosurgical intervention such as deep brain stimulation.

Increasingly, paralleling those laboratory findings that indicated that PD is a diffuse disease affecting much of the central nervous system, is the recognition of the importance of non-motor features of PD. These include a wide array of complaints including sleep disturbance, gait difficulty, autonomic impairment, dysarthria and an array of cognitive complaints ranging from forgetfulness to psychosis. These complaints all require attention and individual treatments; again, ranging from medication for urinary frequency to referral to a physiotherapist for gait training. In many elderly patients with PD, the brunt of the morbidity will be less on motor impairment (fluctuations and dyskinesias) and more on the nonmotor features of the illness.

With respect to the underlying cause of PD, as noted, there have been dramatic breakthroughs, largely brought about by the revolution in molecular biology, not only identifying genes that may cause PD, but more importantly, illuminating the underlying derangements of cellular machinery that are universally found, even in patients without genetic mutations. However, we are still very far from establishing whether PD is entirely genetic or if there are environmental factors implicated. With regard to the genetics, it is important to point out that there is only one moderately common autosomal dominant cause of PD, and this is largely restricted to the Ashkenazi Jewish population. Thus, for the majority of patients with PD who are concerned about the genetic implications of the condition for their families, cautious reassurance is appropriate, bearing in mind that PD is an uncommon disease in the young, and that about 15% of patients with PD have a family history of an affected relative, whereas only 3 - 4 % of healthy septuagenarians report such a history.[7]

Rare tremors

The condition of orthostatic tremor is useful to know about, and its description is pathognomonic. Patients complain of a subjective feeling of unease when standing (only in severe cases when walking). On examination, there may be mild tremor in the arms, and a low amplitude rippling of leg muscles when standing up; patients will tend to stand with wide base, and claw the floor with their toes. The condition typically starts at the age of 65 years, is more common in women, and unfortunately, treatment is unsatisfactory.

Fragile X is the most common genetic cause of mental handicap, and it is now apparent that the male grandparents of these children carry a trinucleotide repeat, which is sufficient to cause neurological deficit as they age, predominantly tremor and ataxia, termed fragile X-associated tremor/ataxia syndrome (FXTAS).[8]

Normal pressure hydrocephalus

As is widely appreciated, this is an important condition characterised by a progressive gait disorder (a gait apraxia), associated with urinary incontinence and cognitive impairment. However, it should be emphasised that this is a rare condition (approximately 1/100 000 per year), and in one series, definite gait improvement was only persistent in a third of patients at 3 years.[9] There is distinct overlap with both cerebrovascular disease and with Alzheimer's disease in many patients, and worsening of cognition and dementia are common in the years following shunting.[10]

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