

Meige's syndrome and bilateral pallidal calcification**J. Herraiz, J. Roquer, D. Escudero, and E. Masó**

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Sirs: The aetiology of Meige's syndrome and essential blepharospasm is unknown. Some neurologists believe that both are focal dystonias of organic origin [5, 6, 8] and others think that these diseases have a psychogenic cause [13].

Recently several papers have argued the organic aetiology of these entities by considering their relation to processes known to involve the extrapyramidal system. These arguments are as follows: the association between blepharospasm and extrapyramidal diseases (parkinsonism, postencephalitic parkinsonism, levodopa and neuroleptic induced dyskinesias, etc...) [3–9]; anatomical lesions demonstrated in the brain stem [1, 4]; and, more recently, by bilateral basal ganglia infarction on CT scanning in a patient with blepharospasm who injected himself with rat poison [6].

Meige's syndrome may also occur in association with such diseases as hypothyroidism and also with autoimmune diseases such as systemic lupus erythematosus, rheumatoid arthritis, Sjögren syndrome and myasthenia [3]. Jankovic and Ford [3] found an increase in antinuclear antibodies titres (more than 1/80) in 15 out of 58 patients in their series.

We report a patient with Meige's syndrome associated with Sjögren syndrome, in whom CT showed bilateral basal ganglia calcification. We believe that this case gives further support for an extrapyramidal cause of Meige's syndrome.

A 60-year-old right-handed woman with a past history of hypertension and diabetes was admitted to hospital for investigation of blepharospasm, which had developed 6 months previously. This was aggravated by bright light, stress and fatigue — a phenomenon more marked at the end of the afternoon. Eventually she had to give up her activities because of a functional blindness caused by the blepharospasm. Neurological examination showed bilateral blepharospasm with increased blinking but without myasthenic reaction to fatigue. No other abnormalities were found. Three months later oromandibular dystonia developed. The following laboratory investigations showed normal results: haemogram, white cell count, urea, creatinine, cholesterol, creatine kinase, serum glutamic oxalo-acetic transaminase, serum glutamic pyruvic transaminase, calcium, phosphorus, sodium, serum protein and serum complement levels. Fasting blood glucose was 132 mg%. Syphilis serology was negative. The Schirmer test was abnormal and parotid scintiscan was compatible with Sjögren syndrome. EMG studies were normal. CT showed bilateral pallidal calcification. The patient was treated with trihexyphenidyl and clonazepam with good effect.

To our knowledge this is the first reported case of an association between Meige's syndrome and bilateral pallidal calcification. Bilateral basal ganglia calcification is a not infrequently found on CT scans. It is usually located in the globus pallidus [2, 11] and frequently is only a radiological

finding without clinical significance. On the other hand, in large series reporting basal ganglia calcification there is nevertheless a certain correlation between this finding and extrapyramidal diseases. Murphy [10] found 3 cases of extrapyramidal disease in 35 patients with bilateral basal ganglia calcification. Sachs et al. [12] found 3 in 14 cases. We found 4 patients with extrapyramidal disease out of 15 cases with bilateral basal ganglia calcification (2 patients with Parkinson's disease, 1 with generalized dystonia and the present case) [11]. Another report of an association between bilateral basal ganglia calcification and Parkinson's disease has been published [7], which suggests that the parkinsonism in such patients tends to be less responsive to levodopa therapy. Only one of our patients with parkinsonism associated with basal ganglia calcification was treated with levodopa, with poor results.

Although it is known that bilateral basal ganglia calcification is frequently a non-specific finding, we believe, on the basis of these reports, that in a proportion of cases this may be related to the development of extrapyramidal disease. Thus the association between Meige's syndrome and bilateral pallidal calcification in our patient may constitute yet another argument supporting the extrapyramidal origin of this condition.

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