

## Small spiral, big mass

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A right handed 44-year-old woman presented with a 4-month history of sudden onset right leg weakness and a 4-week history of small handwriting. On examination, facial expression, blink rate and spontaneous and associative movements were normal. Power in her right leg was mildly reduced proximally with a collapsing, variable nature and she was unable to perform ankle dorsiflexion. Hoover's sign was positive. She had slight impairment of dexterity in the right arm but there was no bradykinesia. The stretch reflexes were normal and symmetrical and the plantar responses down going. On pen and paper tasks she had right sided small script but there was no size decrement, and the Archimedes spiral was small in size with consistent spiral turn spacing. (Figure 1A &B). MRI brain showed a large left parafalcine meningioma with areas of acute/subacute blood internally and intense enhancement with probable infiltration of the superior sagittal sinus at which point the mass crossed the midline. There was no basal ganglia distortion by the mass. (Figure 2) She was managed with a combination of surgical debulking and tapering oral steroid therapy. Her writing and spiral assessments improved over the course of treatment with an initial dramatic improvement noted shortly after the initiation of steroid. (Figure 1C & D)

## Discussion

Although Pick and later Kinnier Wilson (1) considered micrographia, defined as a reduction in size in a patient's handwriting, as probably being of cortical origin, it is largely considered a sign of basal ganglia disease in contemporary practice. While there are a range of reports implicating cortical pathology including masses resulting in parkinsonism, cases resulting in isolated micrographia are rare and include frontal infarction (2) and parietal demyelination. (3) The dramatic improvement with corticosteroids in our patient is presumably due to a resolution in oedema and is similar to the response seen in a case reported with multiple sclerosis. (3) Further support for a cortical origin of micrographia come from post-operative inadvertent 'lesioning' of the dominant fronto-parietal region, stimulation studies implicating Brodmann area 6 and a recent functional imaging study suggesting an aberrance in the network involved in the programming and execution of motor sequences (the

supplementary motor area (SMA), pre-SMA and rostral cingulate motor area) may be responsible. (4)

The correlation between micrographia severity in Parkinson's disease and bradykinesia occurring on finger tapping is not strong and it is possible that when it occurs a disturbance in cortical motor circuitry may also be involved. However the distinctive progressive reduction in the size of the handwriting is what distinguishes it easily from the 'cortical' micrographia seen in this case. The Archimedes spiral drawing in Parkinson's disease is typically asymmetric, and small with spirals compressing further with turns which were not seen in our patient. In right handed patients with Parkinson's disease the script also tends to slope upwards which was not the case in this patient. In patients with pathologically defined Progressive Supranuclear Palsy and in focal pallidal disease 'pallidal handwriting' is occasionally seen in which the script is extremely small but the speed of writing is normal or increased, and there is no decrement in the size of letters.

This case highlights the potential utility of writing and spiral assessments in cases with functional features and few objective motor abnormalities, and the contribution of cortical pathology to micrographia.

Figure legend

Figure 1: Writing and Archimedes spirals pre and post treatment

Figure 2: Images of mass

### **Acknowledgements**

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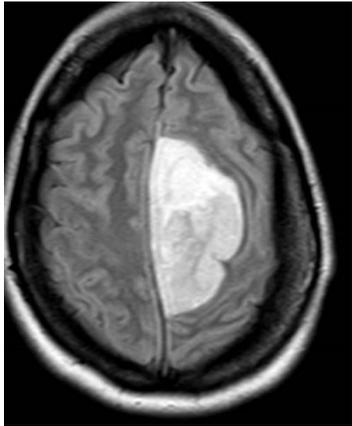


Figure 2A Axial flair demonstrating parasagittal meningioma

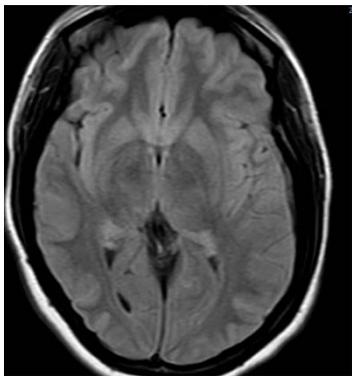


Figure 2B Axial flair demonstrating the sparing of basal ganglia

A 44-year-old woman presented with a 4-month history of sudden onset right leg weakness and a 4-week history of small handwriting. On examination, facial expression, blink rate and spontaneous and associative movements were normal. Power in her right leg was mildly reduced proximally with a collapsing, variable nature and she was unable to perform ankle dorsiflexion. Hoover's sign was positive. She had slight impairment of dexterity in the right arm but there was no bradykinesia. The stretch reflexes were normal and symmetrical and the plantar responses down going. On pen and paper tasks she had right sided small script but there was no size decrement, and the Archimedes spiral was small in size with consistent spiral turn spacing. (Figure 1A, B)

What is her likely diagnosis?

- A. Functional neurological disorder
- B. Parkinson disease
- C. Space occupying lesion (Left hemisphere)
- D. Richardson syndrome

Answer & Explanation:

Answer C. This is a young patient with a stepwise deterioration rather than a progressive one. Her initial symptom was weakness and subsequent one writing difficulty. Although her exam suggested a degree of functional overlay, her writing assessment was suggestive of an organic diagnosis. She did not have other features of Parkinson disease (PD) or Richardson syndrome although this in itself does not exclude the diagnosis. Further support against these diagnoses can be gained from the writing and spiral assessments with PD patients exhibiting decrement and Richardson patients showing abnormalities of writing speed.

#### References:

1. Inzelberg R, Plotnik M, Harpaz NK, et al. Micrographia, much beyond the writer's hand. *Parkinsonism and Related Disorders* 2016; 26: 1-9.