PARANEOPLASTIC PROGRESSIVE-SUPRANUCLEAR PALSY LIKE BRAINSTEM SYNDROME ASSOCIATED WITH LUNG ADENOCARCINOMA

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Objectives Progressive supranuclear palsy (PSP) is a neurodegenerative condition characterised by Parkinsonian features, cervical dystonia and ophthalmoparesis. Paraneoplastic PSP has previously been reported in the literature in association with several different cancer types but is very rare.1–3

Methods Case review.

Results A 74 year old Chinese man was diagnosed with Stage 1b EGFR positive lung adenocarcinoma and underwent a left upper lobectomy. Twelve months later he presented with rapidly progressive neck stiffness, reduction in motor function and gait (over 8 weeks) and recalcitrant disequilibrium. He had hypomimia, frontalis over-activation, blepharospasm, blepharotremor and a supranuclear gaze palsy. There was marked axial rigidity, bradykinesia and cervical dystonia. An MRI brain and spine were unremarkable. Vestibular function tests were normal. Serum antineuronal antibodies were negative. The CSF analysis was unremarkable.

The patient responded to plasma exchange on a two to three-weekly basis with significant improvement in saccadic eye movements and Parkinsonism. However, disequilibrium remained a persistent problem despite the discovery and excision of a second EGFR wild type non-small cell lung cancer, and Rituximab was recently started. Cervical dystonia was treated partially with Botulinum toxin injections, but the patient responded poorly to L-dopa.

Conclusions This suspected paraneoplastic disease exhibits several features of PSP. In particular, the supranuclear palsy, Parkinsonism and dystonia are similar to the typical syndrome. However, the rapidly progressive presentation and disequilibrium are unusual and a response to plasma exchange, suggests humorally mediated neuronal pathology. In rapidly evolving PSP-like cases with cancer, investigation for immunopathology is warranted.

REFERENCES

