A case of paraneoplastic brainstem encephalitis associated with noncutaneous Merkel cell carcinoma

Dr S Hiranput, Dr V Gott
Department of Stroke Medicine, Arrowe Park Hospital, UK

INTRODUCTION

To describe the diagnostic challenge of paraneoplastic neurological syndromes (PNS) in a rare neuroendocrine skin cancer

CASE REPORT

A 70 year old Caucasian male presented with 6 weeks history of dysarthria, dysphagia, and reduced mobility. Neurological examination revealed bilateral facial and bulbar weakness, diplopia in all directions of gaze, and fatiguable upgaze with increasing ptosis and proximal limb weakness. The patient was initially diagnosed as myasthenia gravis. He was given 5 days of intravenous immunoglobulin and 5 sessions of plasma exchange with no improvement. Subsequently, he was investigated for underlying paraneoplastic aetiology. He was found to have positive anti-Hu antibodies and histologically proven Merkel cell carcinoma. The patient has had palliative radiotherapy following specialist skin MDT discussion. He has persistent bifacial and bulbar weakness with restricted eye movement due to chronic innervations. The previous limb weakness has resolved.

INVESTIGATIONS

- MRI head: hypersignal in the pons (figure 1)
- Serum anti-Hu antibodies strongly positive
- CSF: Normal WCC and protein, Anti-Hu band detected
- CT thorax, abdomen and pelvis: no evidence of malignancy
- PET scan: mass in the left thigh/groin (Figure 2)
- Immunohistochemistry of left groin node biopsy suggests Merkel cell carcinoma

Figure 1. T2-axial flair MRI demonstrate signal hypersensitivities in the pons

Figure 2. PET-CT scan shows an increased uptake in left thigh/groin

DISCUSSION

- Approximately 80% of PNS manifest before the diagnosis of the underlying malignancy (1).
- We describe a case of paraneoplastic brainstem encephalitis as a rare clinical presentation of noncutaneous Merkel cell carcinoma, and with positive anti-Hu autoimmune response. Though, in more than half of the reported PNS cases, onconeuronal antibodies are absent (1).
- The lack of sensitive diagnostic test for PNS poses a real challenge in the diagnosis. Since the clinical outcome depends on successful treatment of the malignancy, a thorough investigation for neoplasms is crucial where the syndrome is suspected.

REFERENCES