Case study: Primary Melanocytic Tumours of the Central Nervous System

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Introduction

Primary melanocytic tumours of the central nervous system (CNS) are a group of rare neoplasms arise from the leptomeningeal melanocytes of neural crest origin.1,2 They include meningeal melanocytosis, melanomatosis, meningeal melanocytoma, and meningeal melanoma, varying in aggressiveness, neuroradiological features, morphology, and immunohistochemistry.3 They present primarily in the spinal cord and posterior fossa.2

Case

Ms G, a 67 years old lady presented with a 6-month history of progressive thoracic back pain in 2014. She also suffered from a 1-year history of paraesthesia and weakness of her lower limbs bilaterally. Her back pain also radiated to her legs, leading to a significant reduction in mobility.

She underwent an urgent MRI of the spine with contrast (Fig. 1) which showed an intradural lesion posterior to the T11 vertebral body. (Red arrowhead) MRI Axial (B), the lesion is compressing the spinal cord. (Yellow arrowhead)

The post-operative MRI showed a residual tumour (Fig. 2A), which led to recurrence. Different new enhancing tissues were also identified. (Fig. 2B & C) This diffuse nature is a characteristic of Melanocytosis. Ms G’s case was discussed in the Neuro-Oncology MDT. Radiotherapy was commenced in 2015. CT TAP was also performed to exclude metastatic disease.

Ms G remained stable until 2016 when she presented with worsening neurological deficits due to meningeal melanocytosis at L5 (Fig 2E). Neurosurgery was arranged. However, it was subsequently cancelled due to evidence of cervical spine involvement and potential cerebellar metastases identified on the pre-operative MRI of the spine. The follow up MRI head with contrast showed multiple new lesions in the posterior fossa (Fig 3A & B).

Currently, Ms G is undergoing immunotherapy and will continue to require radiological follow up for treatment progress.

This case inspired me to consider specialty training in Radiology by:

1. Emphasising the importance of radiologist in a multidisciplinary team setting.
2. Exemplifying the duties of radiologist, e.g. tailor MRI sequence according to clinical information (MRI protocols) to provide the best possible imaging results.
3. Demonstrating the importance of attention to details in radiology.
   – The identification of potential brain metastasis in Ms G’s MRI spine helped prevent unnecessary neurosurgery.
4. Exemplifying the radiologist’s ability to recognise different diseases with similar radiological features, e.g. meningioma, lymphoma and nerve sheath tumour.
5. Showcasing the correlation between anatomical abnormalities and clinical manifestations, e.g. spinal cord displacement and compression on MRI.
6. Illustrating the continuity of care in radiology – Ms G has been regularly followed up with MRI spine.
7. Recognising different imaging modalities in monitoring disease progression – CT TAP and Serial MRI spine follow up.
8. Showcasing that radiology is not only for the diagnostic purpose but also has a crucial role in pre-/post-procedural and disease progression monitoring.
9. Exemplifying the opportunity of continuous learning in radiology – to see rare diseases in different specialties.
10. Recognising the workforce shortage in radiology.
   – There were several occasions where Ms G was reviewed in oncological outpatient clinics without the latest imaging reports, which led to delay in clinical decision making.

Fig. 1 - MRI T1-weighted with gadolinium contrast, sagittal (A), showing an intradural lesion posterior to the T11 vertebral body. (Red arrowhead) MRI Axial (B), the lesion is compressing the spinal cord. (Yellow arrowhead)

Fig. 2 - Postoperative MRI spine with contrast, T1-weighted, sagittal (A), showing the residual lesion. (Red arrowhead) Serial MRI spine with contrast (B-E), showing disease progression with larger T10/T11 enhancement (B) (Orange arrowhead), a new leptomeningeal focus at L1 (C) (Yellow arrowhead), post-radiotherapy MRI (D), and new lesion at L5 in 2016 (E) (Green arrowhead).

Fig. 3 – MRI head with contrast, T1 weighted, axial (A & B), showing leptomeningeal enhancement in the right superior cerebellar peduncle, cerebellar vermis and left trigeminal root entry zone. (Red arrowheads)

References