New onset headache caused by histiocytic sarcoma of the spinal cord and leptomeninges: a case report

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ABSTRACT

Background Headache due to raised intracranial pressure is rarely caused by spinal lesions. We describe a patient with primary histiocytic sarcoma who presented with a new onset headache due to raised intracranial pressure and subtle signs of cauda equina syndrome due to predominant lower spinal cord infiltration and minimal intracranial involvement.

Case A previously well 54-year-old man presented with a 2-month history of new onset headache with features of raised intracranial pressure. Progression of lower limb weakness was delayed and mild with diagnostic delay resulting from the primary presentation with headache leading to an initial focus on cerebral pathology. Subsequent investigations revealed a previously unreported presentation of primary histiocytic sarcoma infiltrating the cauda equina causing raised intracranial pressure headache.

Conclusion This case highlights the importance of a broad search in the investigation of new onset raised intracranial pressure headache, including imaging of the lower spinal cord. Primary histiocytic sarcoma should be considered in the differential diagnosis of this rare syndrome.

INTRODUCTION

New onset headache in the setting of papilledema and symptoms of raised intracranial pressure (ICP) raises the possibility of intracranial pathology such as intracerebral mass lesions, parenchymal oedema or disturbances to cerebrospinal fluid (CSF) flow. Headache with such symptoms is rarely described as resulting from lesions in the spinal column.

Intradural spinal infiltrative lesions have been reported with many neuroepithelial cells and other central nervous system (CNS) tumours, spinal nerve tumours, tumour-like cysts and metastatic deposits. Intradural extramedullary infiltration by histiocytic sarcoma, a rare neoplasm arising from histiocytes, a mononuclear phagocytic cell of myeloid lineage has not previously been reported. The typically reported presentation of histiocytic sarcoma is in lymph nodes, skin or gastrointestinal tract with rare primary extranodal presentation in the CNS.

We present a patient with new onset headache with features of raised ICP, who described non-specific systemic symptoms and developed delayed onset lower limb symptoms. The cause of his presentation was detected to be a spinal intradural extramedullary histiocytic sarcoma without significant intracranial lesions.

This case demonstrates the rare presentation of headache attributed to increased ICP resulting from a spinal lesion. In addition, histiocytic sarcoma has not previously been reported presenting as an infiltrative spinal cord lesion, thus expanding the clinical phenotype for this rare neoplasm.

CASE REPORT

A 54-year-old previously well man presented with the complaint of headache worse with recumbency and blurred vision. Symptoms had been present for 2 months and he reported moderate weight loss during this period. Funduscopy revealed papilledema. Shortly after admission, he was noted to have mild strength reduction (Medical Research Council grade 4) in bilateral hip and knee flexor and ankle plantar flexor muscle groups. Reflexes were sluggish in the upper limbs and absent at the knee and ankle joints. The plantar response was flexor bilaterally. Sensory examination was normal in the lower limbs and there was no saddle anaesthesia, anal sphincter tone was normal. On further direct questioning, the patient reported no symptoms of bladder or bowel control.

MRI brain, including the orbits, showed optic disc swelling consistent with papilledema, however, there were no parenchymal lesions. The oculomotor, trigeminal, abducent and facial cranial nerves were thickened and enhanced post-contrast. Lumbar puncture was unsuccessful both at the bedside and subsequently following fluoroscopic guidance. MRI spinal cord undertaken thereafter showed diffuse leptomeningeal enhancement.
Immunohistochemical staining. The cells were positive for leucocyte common antigen on lymphoid morphology negative for keratin and S-100 with a weak positive staining for CD4 and CD163, respectively. Panel H: leucocyte common antigen (LCA) stain was weakly positive. This pattern of staining is in keeping with a diagnosis of histiocytic sarcoma.

Radiological features of histiocytic sarcoma vary depending on the site of involvement and are non-specific. Imaging abnormalities include heparotospleno-megaly and multifocal FDG-avid disease involving lymph nodes. Several reports have demonstrated very intense FDG uptake on PET, particularly in the setting of solid mass lesions, as seen in our patient. Treatment for histiocytic sarcoma is individualised as no standard treatment protocol has been established; though even with treatment, this is an aggressive disease that typically results in death within 2 years.

CONCLUSION

The present case expands the phenotype of extranodal histiocytic sarcoma while also emphasising the importance of a broader search for the aetiology of raised ICP headache in the absence of a causative lesion on primary brain imaging.
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