Findings that shed new light on the possible pathogenesis of a disease or an adverse effect

Gliomatosis presenting as a relative pupil-sparing third nerve palsy in a hypertensive diabetic

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Summary

We present an unusual case of a 41-year-old male patient who presented to the ophthalmology department giving a 3-month history of right sided ptosis, weight loss, diplopia and headache. Clinical examination revealed a right sided relative pupil-sparing third nerve palsy. MRI scan of brain showed thickening of both third nerves. Further investigations revealed a glioblastoma.

BACKGROUND

This case illustrates the importance of urgent neuroimaging in patients presenting with third nerve palsies. While mass lesions giving rise to third nerve palsies are well described, invasion of gliomatosis cerebri into the third nerve with thickening has not to our knowledge been previously reported. This case highlights the need for prompt and appropriate investigation even where other microvascular aetiologies appear more likely.

CASE PRESENTATION

A 41-year-old heavy machinery operator, with a medical history of type 2 diabetes and hypertension, presented to the ophthalmology department. He gave a 3-month history of right sided ptosis, diplopia, headache and weight loss. Systemic and neurological examination was otherwise normal. His medications on presentation included gliclazide, metformin and simvastatin. He lived with his wife. He was a smoker and drank alcohol socially. As he was adopted nothing was known about his family history.

INVESTIGATIONS

Routine blood tests revealed glycosylated haemoglobin of 7.4%. An urgent MRI scan of his brain was requested showing increased signal in the right caudate nucleus and right temporal lobe and bilateral 3rd nerve thickening along with meningeal enhancement (figure 1A,B). MRI of the spinal cord was normal. A right sided relative pupil-sparing third nerve palsy was diagnosed. Other routine blood tests, including biochemical profile, blood count, coagulation screen, thyroid function, vasculitis screen, tumour markers, angiotensin converting enzyme, creatine kinase and inflammatory indices were normal or negative. Neuronal antibodies were negative.

Cerebrospinal fluid (CSF) examination (normal values in parenthesis) revealed a normal opening pressure, glucose and lactate dehydrogenase with a protein of 0.78 g/l (0.2–0.5 g/l), 4 lymphocytes (<5) and 150 red cells. The same

oligoclonal bands were detected in both serum and CSF. The sample was unsuitable for cytological diagnosis. A CT of the thorax, abdomen and pelvis demonstrated a single 4 mm nodule in the peripheral right upper lobe of unknown clinical significance. A repeat MRI brain after 3 weeks demonstrated disease progression, with thickening of both third nerves and adjacent meninges (figure 1C).

DIFFERENTIAL DIAGNOSIS

A wide differential diagnosis, including inflammatory and malignant processes was considered.

OUTCOME AND FOLLOW-UP

Fluorodeoxyglucose (FDG) positron emission tomography (PET) showed a hypermetabolic subependymal lesion in the right hemisphere consistent with malignancy, including lymphoma (figure 1D). Systemic FDG PET was normal. Stereotactic biopsy of the hypermetabolic lesion confirmed a histological diagnosis of glioblastoma, WHO grade 4. If PET scan had not been available, then the area of the brain which appeared most actively involved preferably with enhancement in the least eloquent area would have been biopsied. During radiotherapy planning, he presented with a left relative pupil-sparing third nerve palsy. He did not respond to high dose dexamethasone and was subsequently referred to palliative care.

DISCUSSION

Third nerve palsies are a common presentation, with a wide differential diagnosis, and warrant urgent imaging.¹ A significant proportion of relative pupil-sparing palsies are of unknown cause but are presumed to arise from ischaemia of the vasa nervorum, often within the context of microvascular risk factors such as diabetes or hypertension.² In this context, anisocoria is usually restricted to 1 mm or less and the prognosis is often favourable.³ Lesions of this nature are frequently associated with pain. Clinical variables such as pain or degree of external or pupillary ophthalmoplegia do

Figure 1 (A) Initial transaxial T2 image demonstrating high signal abnormality in the right caudate nucleus. (B) Initial coronal FLAIR section also showing infiltration within the medial aspect of the right temporal lobe. (C) Contrast enhanced T1 MRI 3 weeks after the initial imaging demonstrating bilateral third nerve thickening with meningeal enhancement (arrow). (D) Transaxial FDG PET with a clear area of high glucose uptake within the right caudate nucleus.

not however discriminate between infarction and a mass lesion.^{3 4} If the patient had presented with bilateral pupil-sparing third nerve palsies then the differential diagnosis would have included microvascular, vasculitic, inflammatory, infectious and malignant causes along with polyneuropathies, pseudo-oculomotor deficit, subarachnoid haemorrhage, intracranial hypotension and congenital bilateral oculomotor palsy. While mass lesions giving rise to third nerve palsies are well described, invasion of gliomatosis cerebri into the third nerve with thickening has not to our knowledge been previously reported. This case highlights the need for prompt and appropriate investigation even where other microvascular aetiologies appear more likely.

Learning points

- Urgent neuroimaging (MRI of the brain and magnetic resonance angiography of the head or if not immediately available, CT scan of the head and CT angiography of the head) should be considered in patients presenting with third nerve palsies.
- A wide differential diagnosis should be considered.
- Prompt and appropriate investigations (neuroimaging as described above) should be carried out even where other microvascular aetiologies appear more likely.

Competing interests None.

Patient consent Obtained.

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