Reminder of important clinical lesson

An unusual presentation of an epidermoid brain tumour: a tale of two specialties

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Summary

A 33-year-old right-handed lady was referred to the psychiatry and neurology services by her general practitioner. Previously, she was under psychiatric care for bipolar affective disorder. Recently, her mood had deteriorated prompting the re-referral to the psychiatrists. In addition she had strange attacks. These strange attacks seemed to her like 'sensory overload' or that the 'brain just stops'. Other sensations throughout the attacks included feeling like she is in a 'fish bowl' and surrounding sights and sounds were distorted. She could not speak. After the attack she was hot and flustered, suffered memory loss and was tearful. Both the psychiatrist and the neurologist considered the possibility of these attacks being psychiatric in aetiology. However, the alternative possibility of a coexistence to epilepsy and depression was investigated and MRI demonstrated an epidermoid tumour with the supratentorial portion displacing the left temporal lobe.

BACKGROUND

This case demonstrates an unusual cause of complex partial seizures. Epidermoid tumours make up only 0.8–1.2% of all primary brain tumours, and are rarely found within the middle cranial fossa. The case also underlines the risk of missed or delayed organic diagnosis in patients with a psychiatric illness and bizarre symptoms.

CASE PRESENTATION

A 33-year-old lady with a provisional diagnosis of bipolar affective disorder was referred to the mental health services with low mood. Her initial diagnosis was based on rapid cycling between hypomanic and depressive episodes. Initially, quetiapine 200 mg was introduced but due to intolerance this was changed to fluoxetine. She failed to attend several outpatient appointments but was eventually re-referred for assessment of persistent low mood. She had a significant family history of mental illness. Her three brothers had been diagnosed with depression, schizophrenia and harmful use of illicit substances. She also had a son whom she suspected to suffer from Asperger's syndrome.

It was noted that escitalopram 10 mg once per day had been started in place of fluoxetine for approximately 6 months prior to review. Symptoms were consistent with a moderate depression, poor sleeping pattern, over-eating, low energy and anhedonia. She reported suicidal ideation without intent. There were no features to suggest hypomania or mania. The patient had additionally noted numerous stereotyped episodes of 'déjà vu' over the last 6 months. Episodes lasted approximately 20–30 s followed by a half-an-hour period of confusion and tearfulness.

Her antidepressant therapy was changed to sertraline resulting in a moderate improvement in mood.

A neurology referral was considered, but the psychiatric team instead suggested a period of observation. The patient reported a mild improvement in the frequency of 'déjà vu' episodes at subsequent follow-up appointments, although they were still persistent throughout her time under the psychiatry.

The patient attended her general practitioner (GP) surgery when she suffered a significant worsening of her 'déjà vu' episodes. She was referred to the Mersey Regional Epilepsy Clinic by the GP. Consistent with her previous accounts she reported stereotyped episodes of 'sensory overload' and an unusual familiarity lasting several seconds. She also described a sensation of her head being 'trapped inside a fish bowl,' along with distortion of her immediate surroundings and nominal memory difficulties. Collateral history from her family confirmed stereotyped episodes preceded by silence, followed by confusion without automatisms. Episodes could also occur during 'panic attacks' (without epigastric sensation) and she was often emotionally labile with crying after the event. The frequency was approximately eight episodes per day, each lasting around 30 s, with the majority occurring in the morning.

There was no history of febrile seizures, convulsion, significant head injury, previous central nervous system infection or family history of epilepsy. The systemic and neurological examinations were unremarkable.

INVESTIGATIONS

An outpatient MRI and EEG were booked. The interictal EEG was normal, but during hyperventilation she had a characteristic episode associated with sharp and slow wave activity over the left hemisphere maximal over the left fronto-temporal regions. There were no changes on her ECG during this period. Her MRI demonstrated a

Figure 1 MRI transverse view. Hyperintense signal generated by the epidermoid tumour on T2-weighted imaging.

large lobulated mass measuring approximately 6×2 cm, extending from the left cerebellopontine angle to the medial aspect of the left temporal lobe. There was invagination and displacement of the left hippocampus along with the adjacent brainstem (figure 1). Centrally, within the mass heterogeneous signal was noted on FLAIR (Fluid Attenuated Inversion Recovery) sequences, with associated restricted diffusion. These appearances were consistent with an epidermoid tumour (figure 2).

Figure 2 MRI coronal view. Heterogeneity of signal from the epidermoid tumour is demonstrated on Fluid Attenuated Inversion Recovery sequence with areas of restricted diffusion within the mass. It is possible to see the tumour intruding into the hippocampus and brain stem in this image.

DIFFERENTIAL DIAGNOSIS

- Simple partial seizures
- Complex partial seizures
- Panic attacks
- Cardiac arrhythmia
- Migraine equivalent.

TREATMENT

The patient was started on lamotrigine 100 mg twice daily, clobazam 10 mg once daily, sertraline was discontinued. She had subsequent neurosurgery.

OUTCOME AND FOLLOW-UP

Medication had a modest effect on the seizure frequency. She became seizure-free after surgery. Her last postsurgical follow-up was 3 months after surgery. Her last psychiatric review was 1 month postsurgery, reporting ongoing stabilisation in mood, with no evidence of depression or hypomania. She declined further community psychiatry follow-up.

DISCUSSION

The distinction between epileptic seizures and non-epileptic seizures in the outpatient setting can be difficult. In hindsight the stereotypy, short duration and déjà vu episodes were a strong pointer towards epilepsy. The association 'panic attacks' and depression may or may not have been part of the seizure semiology. A rising epigastric sensation can classically be seen in temporal lobe epilepsy that may also be confused with panic attacks. In this case the imaging proved to be pivotal in reaching the correct diagnosis. Epidermoid tumours are a rare form of an intracranial neoplasm, making up 1.2% of all intracranial tumours.¹ They are more specifically described as inclusion cysts, formed by the implantation of epithelial tissue into another structure.² Epidermoid tumours derive from displaced epithelial remnants from closure of the neural tube between the third and fourth weeks of embryonic development.³ They are extremely slow growing, and usually do not present until the ages of 20–40 years.⁴ Histologically, they are composed of an internal layer of stratified squamous epithelium, covered by an external capsule. Growth is driven by desquamation.⁵ Rarely can epidermoid tumours transform into malignant squamous cell carcinomas.⁶ The most common locations for these tumours are the cerebellopontine angle cistern (40–50% of cases), the parasellar region (30% of cases) and more rarely the middle cranial fossa and spinal canal.⁷ Intraparenchymal locations, as in the case of this patient, are uncommon.⁸ MRI is the imaging modality of choice, specifically with the addition of diffusion-weighted imaging. This helps to aid the distinction between epidermoid and arachnoid cysts. The former demonstrates restricted diffusion, and thus has high signal intensity on diffusion-weighted imaging. The latter appears hypointense secondary to its free water content.⁹ Complete resection is often impractical given the risks of damage to nearby healthy tissue. Due to the generally slow regrowth of tumour remnants, subtotal resection offers a good prognosis.¹⁰ There are a few case reports of epidermoid tumours in the temporal lobe presenting with seizures.^{11–13} Both partial and generalised seizures were reported. The chronological association of the seizures started of with escitalopram is interesting and may have been due to effects on seizure

threshold. With regard to drug treatment we used lamotrigine as it is an effective and tolerable antiepileptic drug with proven efficacy in partial epilepsy.¹⁴ Additionally, it has mood-stabilising effects reducing depressive episodes in bipolar disorder.¹⁵ Given the refractory nature of the patients' seizures and the location of the epidermoid tumour she underwent cytoreductive surgery.

Our case report highlights the complexities of making a diagnosis of epilepsy in the context of psychiatric disease. Features that help one to suspect non-epileptic attacks include long duration and fluctuating course, asynchronous movement, pelvic thrusting, side-to-side head or body movement, closed eyes, ictal crying and good memory recall.¹⁶ Postictal crying should not mislead doctors to assume a non-organic nature of the attacks. Guidance for pursuing organic causes of psychiatric symptoms varies. Currently, neuroimaging in first episode psychosis is not generally advised.¹⁷ Reviewing the literature, a number of cases have been reported with epilepsy and/or brain tumours presenting with psychiatric symptoms that were otherwise neurologically silent.^{18–20} Admittedly the vast majority of patients presenting to psychiatry will not have an organic cause to their presentation. However, the treatable nature and potential consequences of late or missed diagnoses of brain tumours and epilepsy warrants vigilance.

Learning points

- ▶ Epidermoid tumours are a histologically benign slow-growing rare primary brain tumour. They rarely undergo malignant transformation and prognosis in most cases is good.
- ▶ Brief recurrent stereotyped events should raise the possibility of partial seizures especially when associated with other characteristic aura such as déjà vu and postictal confusion.
- ▶ A psychiatric diagnosis should not preclude suspicion of concurrent medical pathology, whether it is neurological or otherwise.
- ▶ Neuropsychiatric symptoms that do not classically fit a psychiatric or neurological explanation may benefit from a multidisciplinary approach to diagnosis.

Competing interests None.

Patient consent Obtained.

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