Paraneoplastic limbic encephalitis with anti-Ma2 antibodies; a rare presentation of differentiated testicular teratoma

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BACKGROUND

Testicular cancer is the most common malignancy in men aged 15-44 years old, with its incidence increasing over the last two decades.

Germ Cell Tumour's (GCT) can be classified as Germ cell neoplasia in situ (GCNIS) and also seminomatous germ cell tumours (SGCT) and non-seminomatous germ cell tumours (NSGCT). Testicular teratoma is classed as a GCT and can be found within one or more of the germinal layers. [1]

Paraneoplastic limbic encephalitis is a rare complication associated with certain malignancies. It presents with features such as memory loss, visual disturbance, seizures and personality changes.[2]

There have been rare cases of paraneoplastic encephalitis reported in well differentiated teratoma with other histological components such as seminoma.[3] More frequently, limbic encephalitis is reported in females with ovarian germ cell tumours.[4]

We highlight a rare case of 100% differentiated teratoma in a male presenting with paraneoplastic limbic encephalitis.

IMAGING

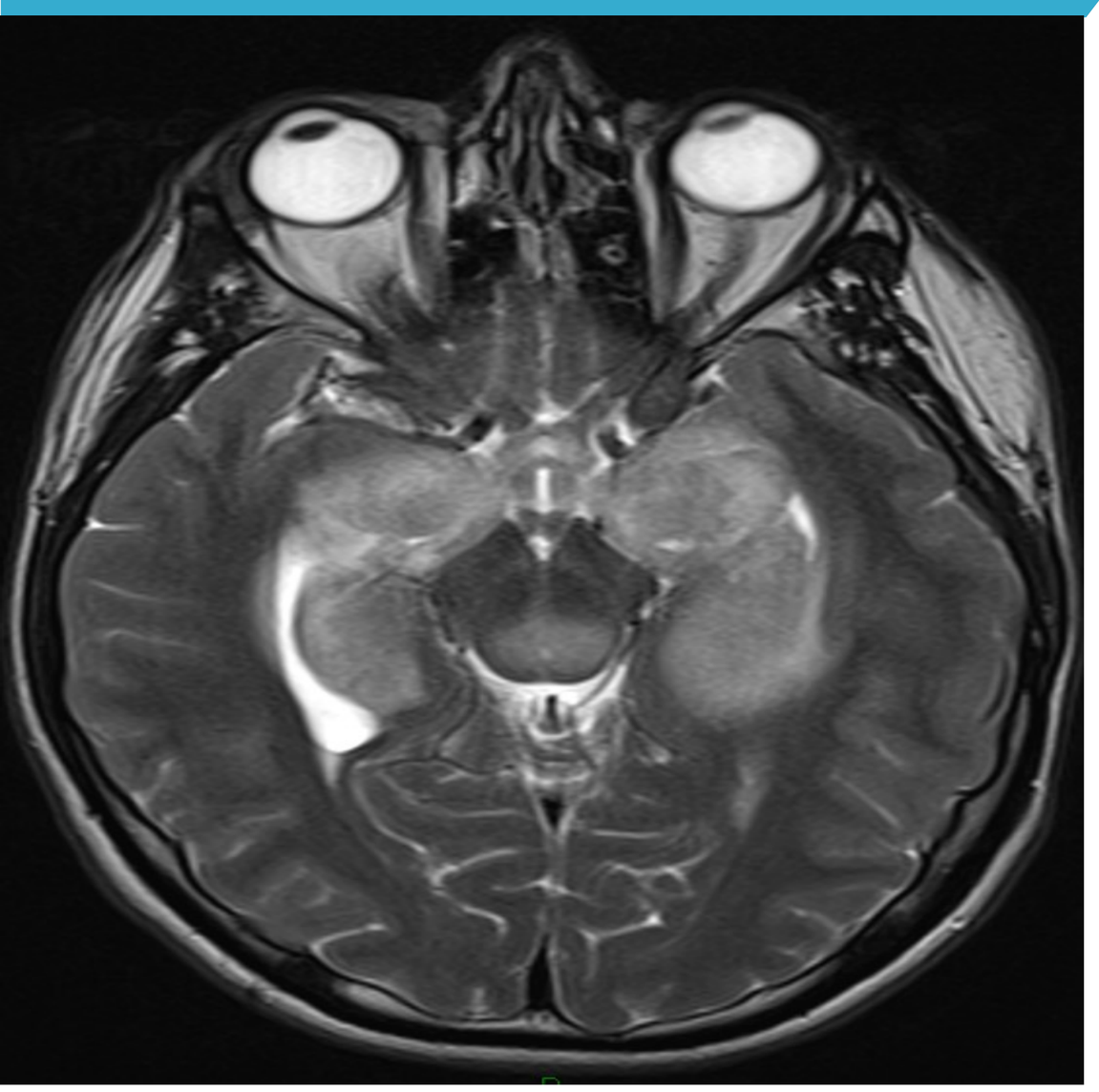

Figure 1. Initial MRI scan – August 2021
Progressive enlargement of bilateral hippocampi with associated high T2 signal. He also had T2 hyperintensities in brainstem. Features consistent limbic encephalitis

CASE PRESENTATION

A 21-year-old gentleman presented with focal seizures, with secondary generalised tonic-clonic seizure. He also had rapidly progressive amnesia with diplopia. After a normal Computed Tomography (CT) head he was discharged with a referral to the first seizure clinic. He was later admitted with seizure activity and an Magnetic Resonance Imaging (MRI) revealed bilateral medial temporal lobe hyperintensities.

An encephalitis screen showed positive Anti-Ma2 antibodies and an ultrasound revealed a right testicular tumour. Tumour markers were negative and a staging CT-Chest/Abdomen/Pelvis revealed no evidence of metastatic disease. Further MRI imaging revealed progressive enlargement of bilateral hippocampi; indicating limbic encephalitis.

Following an orchidectomy he was transferred to a tertiary centre where he underwent plasma exchange. Pathology indicated that it was 100% teratoma post-pubertal type, with mature differentiated cartilage, adipose and muscle tissue. Repeat MRI showed initial response to plasma exchange and high dose prednisolone.

He was re-admitted and required a prolonged hospital stay. His admission was complicated by seizures and aspiration pneumonia. He was initiated on cyclophosphamide and later azathioprine because of worsening features on MRI.

Following discharge, he remained in a residential house for acquired brain injuries and has gradually experienced functional improvements. There has been no evidence of malignancy on his repeat imaging organised by the oncology team in the 20 months since first presentation.

CONCLUSIONS

Anti-Ma2 antibody encephalitis is associated with malignancy in over 90% of cases; including GCT's.[5] This case highlights the importance of screening for malignancy with this diagnosis.

One of the key findings is that pathology revealed purely differentiated teratoma which is rarely reported as causing limbic encephalitis. There have been reports of mixed GCT's presenting with encephalitis, especially in females.[4]

A further important aspect of this case is that the clinical phenotype suggested a characteristic limbic encephalitis with brainstem involvement. This resulted in the neurology team identifying the tumour before the antibodies were available. Consequently, the tumour was treated more promptly. Given the significant morbidity associated with limbic encephalitis, the importance of early diagnosis and treatment is vital.

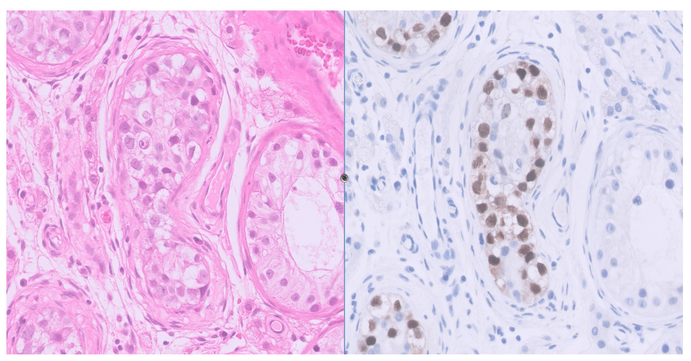

Figure 2. Background tubules with germ cell neoplasia in situ. H&E (left) showing large atypical cells, positive on immunostainins for OCT3/4 (right).

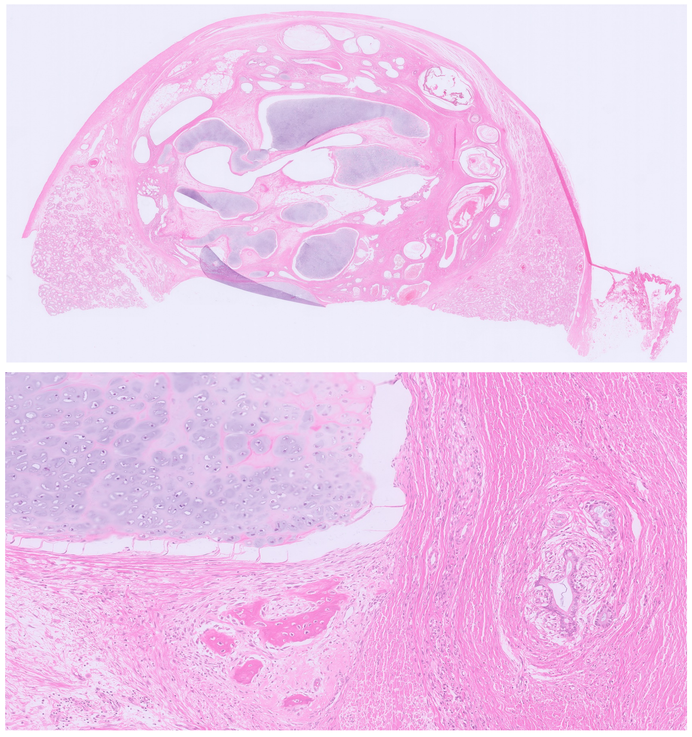

Figure 3. Low power image of testis, showing blue-grey staining nodules of cartilage in the centre, pale fat and epithelial lined cysts at the periphery

Figure 4. Higher power showing mature cartilate (top left), bone (lower left) and glandular epithelium (right).

Images of patient's pathology kindly provided by Dr Jonathon Salmond as referenced above.

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