

'Papaya Seed' Appearance of Subependymal Nodules on Magnetic Resonance Imaging in a Patient with Tuberous Sclerosis

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ظهور عقيدات البطانة العصبية على شكل 'بذور ثمرة البابايا' بتصوير الرنين المغناطيسي في مرضى التصلب الجلدي

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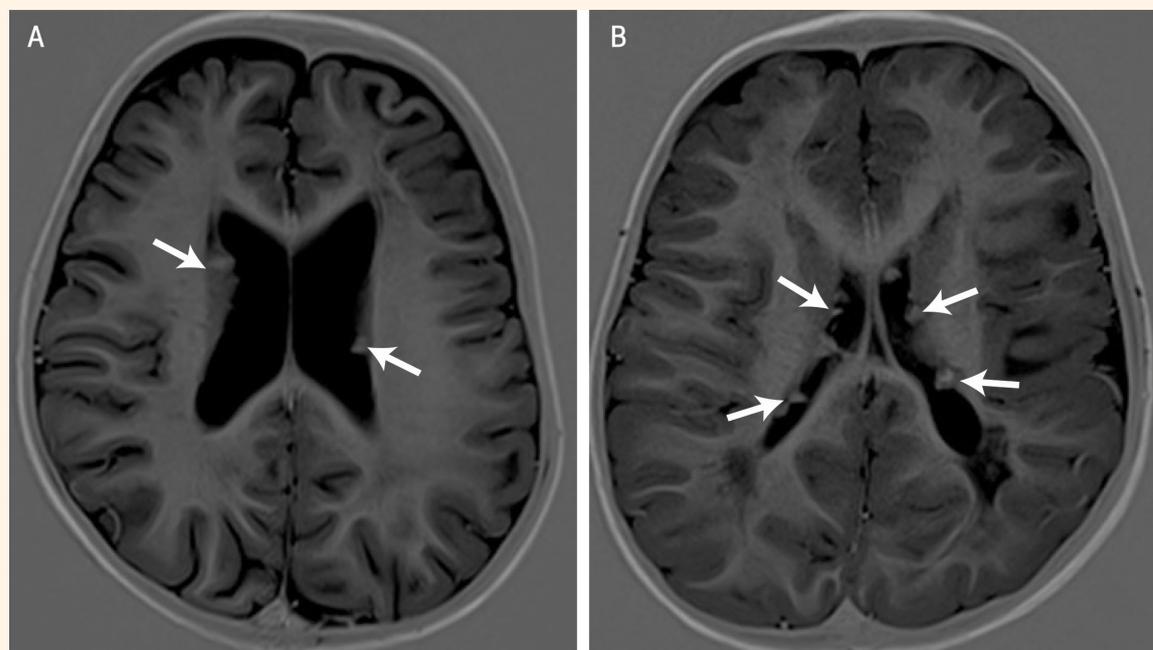


Figure 1A & B: T1-weighted axial magnetic resonance images showing subependymal nodules (arrows) with a 'papaya seed' appearance in two patients with tuberous sclerosis.

A 14-MONTH-OLD MALE WAS REFERRED TO the Department of Child Health at the Sultan Qaboos University Hospital (SQUH), Muscat, Oman, in 2014 with a two-week history of frequent abnormal movements of the body. A video, recorded by the patient's father, revealed that the patient suffered from typical *flexor* body spasms and left side partial motor seizures. On physical examination, the patient had several cutaneous features of tuberous sclerosis, including depigmented skin lesions and Shagreen patches. A neurological examination was

unremarkable. Sleep electroencephalography displayed interictal generalised and focal discharges from the left frontocentral region. There was no evidence of hypsarrhythmia. Magnetic resonance imaging (MRI) of the brain revealed multifocal cortical and subcortical tubers with subependymal nodules (SEN) which had a 'papaya seed' appearance [Figure 1A]. Four to five rhabdomyomas in both ventricles were visible on echocardiography. Ophthalmological examination revealed an astrocytoma along the inferior vascular arcade close to the optic nerve of the left eye. The

patient's father had a possible skin lesion on his back, while the rest of the family had normal examinations. The child was diagnosed with tuberous sclerosis and epilepsy. He was initially prescribed the antiepileptic medication vigabatrin; due to a lack of response, this was changed to topiramate and levetiracetam after two weeks.

Comment

Tuberous sclerosis is a neurocutaneous disease characterised by skin lesions, often in association with epilepsy (80–90% of cases) and mental retardation (40–70% of cases).^{1,2} The disease may affect any organ of the body, but predominantly involves the brain. The diagnostic criteria of tuberous sclerosis have recently been revised with the new criteria emphasising clinical elements of the disease based on 11 major and nine minor features.³ For a diagnosis of definite tuberous sclerosis, evidence of two major features or one major feature with two or more minor features is essential. The presence of one major feature or two minor features is labelled as possible tuberous sclerosis.³ Subependymal giant cell astrocytoma and SEN are considered two major features; the latter are seen in 80% of patients with tuberous sclerosis and can be detected antenatally or at birth.⁴ These benign growths develop along the ependymal lining of the lateral and third ventricles.

Computed tomography (CT) of the brain allows easy visualisation of SEN due to calcification.⁵ An MRI scan alone can easily miss calcific lesions and hence a CT scan is also often performed to confirm the calcification.⁶ At SQUH, analysis of CT and MRI

scans performed on children with tuberous sclerosis has shown that MRI imaging of multiple SEN attached to the ventricular wall closely resemble papaya seeds attached to the interior of the fruit [Figure 1B]. Observations of clinical features of tuberous sclerosis combined with the 'papaya seed' appearance of SEN on an MRI scan could obviate the need for additional CT imaging among patients with tuberous sclerosis.

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