Schizencephaly

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A 71-year-old lady presented to the acute medical unit with dizziness and collapse at home with no loss of conscious. There was no complaint of headache, palpitations or chest pain and there was no evidence of seizures. She had a background of hypertension and hypothyroidism and was on regular bendroflumathiazide and bisoprolol.

On examination her pulse was regular and she had significant postural hypotension on measuring lying and standing blood pressure. It was noted that she has unequal pupils but the rest of her examination was unremarkable. She had a normal ECG and chest X-ray. Routine blood tests were normal as well.

Bendroflumethiazide was stopped and a CT head scan was performed (Figure 1).

Figure 1. The CT scan
The CT scan shows schizencephaly. This is an incidental congenital defect in the brain and it was unlikely to account for the patient’s presentation. The skull frontal bony deformity confirms that this is a developmental defect (personal communication, hospital colleague, 2014).

The cause of the collapse in this case was postural hypotension secondary to diuretic use. The patient improved after stopping bendroflumethiazide.

This case was discussed with the neurosurgical team who confirmed the diagnosis of shizencephaly that does not require any intervention.

The total prevalence of schizencephaly is 1.48/100,000 births.1

Schizencephaly is a rare disorder of neuronal migration in which there are one or more fluid-filled clefts in the cerebral hemisphere that communicate with the lateral ventricle.

Two types of schizencephaly have been described:

- Type 1 has small symmetrical clefts and the edges of the clefts are fused within a pia-ependymal seam that is continuous with the ependyma of the lateral ventricle.
- Type 2 has extensive clefts that extend from the ventricle to the surface of the brain and subarachnoid space and the edges are not fused.2

Children with closed-lip schizencephaly present with hemiparesis or motor delay whereas patients with open-lip schizencephaly present with hydrocephalus or seizures.3

Treatment for individuals with schizencephaly generally consists of physical therapy, treatment for seizures, and, in cases that are complicated by hydrocephalus, a shunt.

The prognosis for individuals with schizencephaly varies depending on the size of the clefts and the degree of neurological deficit.4

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Arachnoid cysts are collections of cerebrospinal fluid within the layers of the arachnoid membrane; the cyst may or may not communicate with the subarachnoid space.

Arachnoid cysts do not communicate with the lateral ventricles.5

The patient in this case was totally asymptomatic and the presentation on this occasion was secondary to postural hypotension.

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References:


