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Severe Hydrocephalus due to Obstructive Basilar Dolichoectasia of the Third Ventricle

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ABSTRACT

Vertebro-basilar dolichoectasia (VBD) is a rare pathology of unknown aetiology. Its clinical presentation is wide and prognosis is generally poor with a high mortality rate. Cerebral magnetic resonance imaging is the gold standard for diagnosis.

We report an unusual case of intracranial dolichoectasia. VBD was revealed during investigation of a patient with altered mental status. CT brain imaging demonstrated severe obstructive hydrocephalus secondary to compression of the third ventricle. Management is always challenging and depends on the location and the mode of presentation. Our patient died despite surgical management with placement of an external ventricular shunt.

LEARNING POINTS

- Vertebro-basilar dolichoectasia is a little known cause of altered mental status in elderly patients.
- An atypical presentation of vertebro-basilar dolichoectasia can mimic extensive cerebrovascular haemorrhage.
- The prognosis is poor despite prompt diagnosis and the surgical treatment of choice.

KEYWORDS

Dolichoectasia hydrocephalus, brain magnetic resonance imaging, vertebro-basilar dolichoectasia

INTRODUCTION

Vertebro-basilar dolichoectasia (VBD) usually presents with few or no symptoms. However, it can also manifest with ischaemic or haemorrhagic phenomena caused by compression of the cranial nerves or, more rarely, with hydrocephalus due to compression of the third ventricle.

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CASE DESCRIPTION

A 73-year-old patient was admitted to the emergency department for sudden onset of altered mental status and confusion. His son reported he had been his usual self earlier in the day. His history included arterial hypertension, hypercholesterolaemia, atrial fibrillation and myocardial infarction. No previous cognitive impairment or functional dependence was reported.

On admission to the emergency room, the patient's Glasgow score was 10/15 (E2V4M6), blood pressure was 190/90 mmHg, heart rate was 65 beats per minute, and saturation was 94% on room air. He was apyretic and capillary glucose was 220 mg/dl.

Complete blood investigations including blood and urine toxicology, were normal. Subsequently, the patient experienced deterioration in consciousness (Glasgow score of 6/15 (E1V1M4)) with bilateral mydriasis, requiring immediate endotracheal intubation and mechanical ventilation. The initial presumptive diagnosis was an extensive cerebral haemorrhage, however, the brain CT scan revealed basilar dolichoectasia that ascended to the third ventricle. This obstructed the foramen of Monro and was responsible for biventricular hydrocephalus with intracranial hypertension *(Figs. 1 and 2*).

The patient was immediately transferred to the operating room and a right external ventricular shunt was placed. The postoperative magnetic resonance imaging (MRI) scan showed that the external ventricular shunt was correctly placed and revealed non-communicating hydrocephalus due to probable extrinsic compression of the floor of the third ventricle by dolichoectasia of the basilar stem (*Fig. 2*).

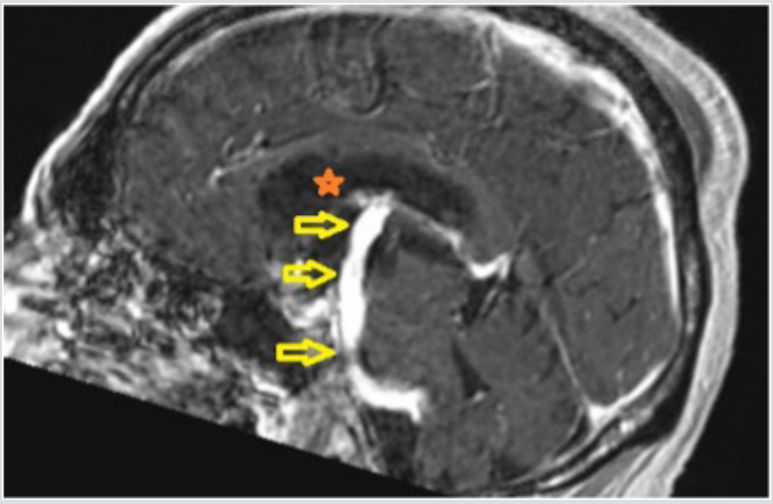

Figure 1. Time-of-flight brain MRI (coronal view) with contrast agent showing dolichoectasia of the basilar artery (yellow arrows) causing dilatation of the ventricular system (orange star)

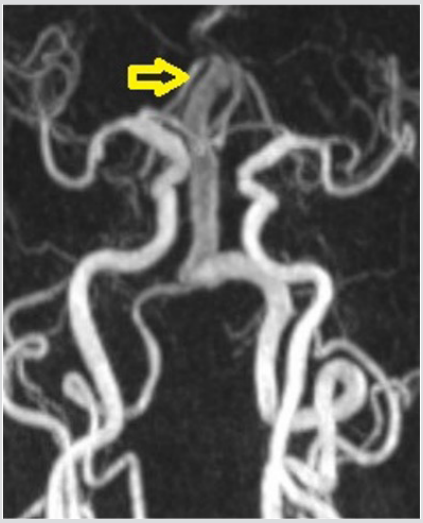

Figure 2- Brain MRI (T1-weighted image with contrast, sagittal view) showing dolichoectasia of the basilar artery (yellow arrow)

No clinical improvement was achieved despite continuous cerebrospinal fluid drainage and optimal management of cerebral oedema. After 3 days in the intensive care unit, the diagnosis of brain death was confirmed based on evoked potentials and a pathognomonic electroencephalogram. The patient died 4 days after admission.

DISCUSSION

VBD is defined by an increase in the length and diameter of one or more intracranial arteries [1]. It is a rare pathology of unknown aetiology but seems to have a genetic component [1].

Clinical presentation is variable. VBD is usually asymptomatic or may present with acute ischaemic phenomena (resulting from local thrombosis or embolism), haemorrhage following rupture, or chronic compression of surrounding structures (brainstem, cranial nerves, or the third ventricle with hydrocephalus) [1, 2]. The reported case presented with an atypical evolution of arterial dolichoectasia.

The frequency of this condition is difficult to estimate. It is associated with approximately 3% of all cerebral infarctions, giving an incidence of 2500 cases annually in France [2, 3]. Some vascular risk factors have been associated with this entity such as age, male sex, hypertension, and a history of myocardial infarction. All of these factors were present in our patient.

Brain CT scanning, cerebral MRI or cerebral arteriography are helpful for diagnosis [3, 4]. A cerebral CT scan without contrast can show hyperdensities corresponding with calcifications in the wall of the dolichoectasia and can also help to identify the mechanisms responsible

for the clinical signs, for example compression of the brainstem or the third ventricle, as in the case presented.

MRI associated with angiographic visualization of the vessels is the gold standard for the diagnosis of intracranial dolichoectasia. It allows good visualization of the arterial lumen and investigation of the relationship between the dolichoectasia and surrounding structures (cranial nerves, brain stem, third ventricle). The sensitivity is superior to that of CT scanning [5]. In contrast, arteriography is used less and less because of the risk of vertebra-basilar ischaemia.

There is currently no established protocol for the management of intracranial arterial dolichoectasia. Therapeutic management (surgical, endovascular, medical or even no treatment) depends on the presentation and location. Dolichoectasia is a therapeutic challenge as it can cause compression, rupture with meningeal haemorrhage, and thromboembolism with cerebral infarction [6].

External ventricular shunt placement is the treatment of choice leading to a better prognosis in some cases, though not unfortunately in ours. In our case, surgery was carried out but with no clinical response.

CONCLUSION

Vertebral dolichoectasia is a rare pathology and can cause compression, thrombosis or rupture, which sometimes makes management difficult. The prognosis depends on the mode of presentation and location. Prognosis is poor despite surgery.

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