Case Report

Foix-Alajouanine Syndrome: Report of a case with spinal cord haemorrhage induced by normal vaginal delivery

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Abstract

Foix and Alajouanine first described the syndrome in two young men with spinal arteriovenous malformation. It was initially thought to be a subacute myelopathy produced by a thrombotic process of the spinal cord that ultimately caused death. In this paper, we report for the first time, the occurrence of intramedullary hemorrhage after a normal vaginal delivery in a 32-year-old primigravida with Foix-Alajouanine syndrome. During pregnancy the patient received subcutaneous heparin, 0.6 ml twice a day until delivery, for prevention of deep vein thrombosis. Our data, for the first time, document objectively the risk of hemorrhagic transformation (presenting with acute compressive myelopathy) posed by normal vaginal delivery in patients with Foix-Alajouanine syndrome.

Key words:
Foix Alajouanine syndrome, spinal arteriovenous malformation, vaginal delivery

Introduction

Foix and Alajouanine first described the syndrome in two young men in 1926 [1]. It was thought to be a subacute myelopathy produced by a thrombotic process of the spinal cord that ultimately caused death [2]. At autopsy, they discovered necrosis of the spinal cord and numerous thickened tortuous vessels lying on the surface of the cord. In 1931, Lhermitte et al. associated this syndrome with spinal arteriovenous malformation (AVM) [3] and attributed the myelopathy to thrombosis of the abnormal vessels within the spinal canal. Over the years, several diagnostic rubrics have been used interchangeably with Foix-Alajouanine syndrome (FAS) including angiodysgenetic necrotizing myelopathy, subacute necrotizing myelopathy, and venous congestive myelopathy [4]. The syndrome is generally believed to be irreversible with a poor prognosis [5,6]. In most patients with this condition, sensory symptoms and leg weakness are the most common initial symptoms [7-9].

Case presentation

A 32-year-old primigravid Saudi female came to the emergency room of King Khalid Hospital on Feb 10, 2012 with a chief complaint of weakness of both legs and inability to void shortly after a normal vaginal delivery. Medical history was significant in that she had developed deep vein thrombosis of the left lower extremity in the first trimester of pregnancy, and for which she was started on low molecular weight heparin (Enoxaparin) 0.6 ml subcutaneously twice a day which was continued until
delivery. On examination the patient was conscious and alert. Her vital signs were as follows: blood pressure 111/76 mmHg, temperature 36.6 °C, pulse rate 92 per minute, respiration rate 12 per minute. Oxygen saturation was 94%. The pre-labor obstetric examination prior to presentation to King Khaled Hospital had shown the uterus to be term size with a normal tone and a conceptional age of 38 weeks. A regular fetal heart rate of 150 beats per minute was present. Initially, vital signs were normal and there were no pallor, icterus or pedal edema. No skin rash or petechiae were present. Her routine blood tests and coagulation profile were normal. She delivered a 3.5 kg baby with an Apgar score of 7/10 and 9/10 in one and five minutes, respectively. However, under observation, she deteriorated neurologically, prompting her urgent transfer to King Khaled Hospital.

General physical examination was unremarkable at admission except for a distended bladder. Neurological examination showed a normal mental status. Cranial nerves were intact. Evaluation of the motor system showed normal strength in the upper extremities. She had flaccid paraplegia (0/5). Deep tendon reflexes were +2 in the upper extremities and absent in the lower extremities. There was no Babinski sign. Cerebellar testing was normal. She had an L4 pinprick sensory level bilaterally. There were no cranial or spinal bruits. There was a moderate nuchal rigidity. Laboratory tests showed a prothrombin time of 11.8, partial thromboplastin time of 22.6, international normalized ratio of 1 and platelet count of 247,000 per μl. Magnetic resonance imaging of thoracic spine showed intraspinal-dural arteriovenous malformation with extramedullary and intramedullary components associated with cord haemorrhage (abnormal bright signal in the spinal cord at T5 level) (Figure 1). There was swelling of the cord from T5 to T10 level with multiple serpentine signal void structures presenting as heterogeneous T1 and T2 hyperintense configurations pointing towards the hemorrhage within the cord (Figure 2). The patient was subsequently transferred to a tertiary-care facility for neurosurgical intervention. Later, she was lost to follow-up.

Discussion

The principal mechanism of progressive myelopathy in FAS is believed to be vascular thrombosis that leads to multiple small infarcts, which can be hemorrhagic in cases of venous thrombosis [10]. Other complications of FAS include subarachnoid hemorrhage, epidural hematoma, hematoma, and compressive myelopathy. In our patient, the presence of nuchal rigidity was consistent with the extension of the cord hemorrhage into the subarachnoid space. It should be noted that Valsalva (bearing down during labor) maneuver may be a contributing factor in the pathogenesis of hemorrhagic complication in FAS, by raising the spinal venous pressure. Similarly, aneurysmal subarachnoid haemorrhage that occurs in adulthood is often temporally related to a Valsalva maneuver (e.g., physical exertion, sexual activity, bowel movements, NVD). It should be noted that AVM can be a culprit behind “thunderclap” headaches associated with neurologic signs, commonly seen in cerebral aneurism. AVM most commonly presents in young adults, and typically occurs within the brain parenchyma, causing intracerebral hematomas (also known as parenchymal hematomas) [11]. Occasionally, these lesions expand into the ventricular or subarachnoid space and produce meningeal signs. Non-complicated AVMs most commonly present with seizures and chronic headaches. In our patient hemorrhagic transformation of AVM occurred
from T5 to T10, presenting with compressive myelopathy.

At this juncture, we would like to alert obstetricians to the risk of hemorrhagic transformation of AVM during vaginal delivery and a possible need for elective cesarean section. Further studies will be necessary to identify various risk factors for hemorrhagic transformation in FAS. In conclusion, spinal arteriovenous malformation is associated with what has been known as Foix-Alajouanine syndrome (FAS), presenting with a clinical picture of sub-acute progressive myelopathy. We caution obstetricians against vaginal delivery in patients with FAS because of a risk of hemorrhagic transformation and abrupt onset of compressive myelopathy.

Conflict of Interest
Authors declare no conflict of interest