Case Report

Acute ischemic stroke secondary to ventriculoperitoneal shunt dysfunction in a child with Moyamoya syndrome

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ABSTRACT

Background: Patients with brain vascular disease and hydrocephalus may be predisposed to acute ischemic stroke in case of shunt dysfunction and subsequent increased intracranial pressure. Patients with brain tumor may develop hydrocephalus as a consequence of obstruction of cerebrospinal fluid pathways and radiation-induced moyamoya syndrome secondary (RIMS) to radiotherapy (RT).

Case Description: A 15-year-old male patient, affected by hydrocephalus and RIMS, presented acute cerebral ischemia after an episode of shunt malfunction. The shunt was promptly revised and the areas of ischemia visible at magnetic resonance imaging significantly decreased.

Conclusion: Children who receive RT for brain tumor, particularly if the circle of Willis region is involved, require close surveillance for the development of vasculopathy and consequent stroke. This surveillance must be even tighter if the patient has been treated with ventricular shunt for the possible synergistic interaction between the two causes on reducing cerebral perfusion and increasing the risk of acute ischemic events.

Keywords: Radiation-induced moyamoya syndrome, Transient ischemic attack, Ventriculoperitoneal shunt

INTRODUCTION

Coexistence of brain vascular disease and hydrocephalus treated with ventriculoperitoneal shunt may predispose to acute ischemic stroke in case of shunt dysfunction and subsequent increased intracranial pressure (ICP). Increased ICP, further, decreases cerebral perfusion pressure (CPP) that is already impaired in patients with vasculopathy. This is a common phenomenon in patients with critical disease (traumatic brain injury and subarachnoid hemorrhage) and lost of vascular autoregulation.

The association of vasculopathy and hydrocephalus is not so uncommon: patients with suprasellar tumor treated by surgery and adjuvant radiotherapy (RT) may develop hydrocephalus as a consequence of obstruction of cerebrospinal fluid (CSF) pathways, and radiation-induced moyamoya syndrome (RIMS) secondary to RT.[3]

Surprisingly, to the best of our knowledge, no cases of acute ischemic stroke at time of shunt dysfunction in patient with cerebral vasculopathy have been reported in the literature.
We, herein, report a case of a 15-year-old male patient with a history of hydrocephalus secondary to optic pathway gliomas (OPG) and RIMS who developed acute cerebral ischemia caused by shunt dysfunction, with secondary partial resolution of ischemic areas after normalization of ICP.

CASE DESCRIPTION

This patient, at the age of 1 year, had a diagnosis of pilocytic astrocytoma of optic pathway that was initially treated with surgical debulking and chemotherapy (CT), according to SIOP 2004 LGG protocol. Due to tumor progression, over the next 3 years, he underwent several surgical procedures and different CT regimens (Carboplatin/Vincristine; Cisplatinum/Vincristine/Cyclophosphamide; and Bevacizumab/Irinotecan). He also developed a hydrocephalus treated with ventriculoatrial shunt at the age of 1 year transformed in ventriculoperitoneal shunt after 6 years. The tumor progressed under chemo so at age of 4 he underwent surgical debulking followed by fractionated RT with a total dose of 54 Gy. One year after RT, he presented multiple episodes of transient ischemic attacks with the right facial palsy and aphasia. Magnetic resonance imaging (MRI), MRI angiography, computed tomography angiography, and digital subtraction arteriography highlighted a tight stenosis of the supraclinoid internal carotid artery (ICA), M1, and A1 segments on the left side, with a contralateral hypoplastic A1 [Figure 1]. A brain positron emission tomography (PET) scan (H2150-W ATER PET) defined an initial stage of RIMS. He started antiplatelets therapy (acetylsalicylic acid 100 mg/day) and underwent a left encephalo-duro-arterio-myo-synangiosis (EDAMS) with multiple burr-hole surgery (MBHS). His final neurological status was characterized by minimal right-side hemiparesis, mild neurodevelopmental delay, and severe visual impairment.

At the age of 15, he was readmitted with a 4-day history of headache and progressive depressed level of consciousness. At admission, clinical condition quickly worsened and he was transferred in intensive care unit with a Glasgow Coma Scale 3/15 and mydriatic pupils. MRI showed increment of ventricular size with restricted diffusion areas on diffusion-weighted images of distal territory of ICAs and left middle cerebral artery (MCA), compatible with acute ischemic injury [Figure 2]. The shunt was revised in emergency. The valve and the ventricular catheter were found to be blocked and were changed. The patient was admitted in intensive care unit and was in neuroprotection for 7 days. At weaning from anesthesia, a right-side hemiplegia was evident. Postoperative MRIs showed progressive reduction of extension and conspicuity of restricted areas on DWI and new-onset small ischemic lesions in basal ganglia, in territory of lenticulostriate arteries of MCA. The patients had a slow recovery until the restoration of his usual clinical conditions after 3 months of rehabilitation.

DISCUSSION

RT is an effective therapeutic modality for the treatment of many pediatric brain tumors, playing a role in local disease control and improving patients’ overall survival, especially those patients who do not respond to CT.[11] Nevertheless, cranial irradiation is the major risk factor for late cerebrovascular complications (LCC) in pediatric population due to iatrogenic damages of cerebrovascular tissue surrounding the radiation target.[11] These are defined as lesions secondary to small, medium, or large vessel abnormality occurring after RT within the irradiated volume. LCCs include microbleeds, cavernomas, and superficial siderosis. Passos et al.[10] reported an incidence of 36% of LCCs in a cohort of 121 children treated with RT for central nervous system tumors, in which the most frequent complications were microbleeds (80.6%) and cavernomas (52.8%). In his series, only one patient developed a moyamoya syndrome with stroke.

RIMS develops particularly in patients with tumors in close proximity to the circle of Willis.[12] As reported by Scala et al.,[11] up to date, 94 cases are so far described in the literature being the incidence highest in patients with OPG.

Pathogenesis of RIMS is debated in the literature: several authors support the theory of a chronic arteritis obliterans caused by the formation of reactive oxygen species that provoke reactive proliferation of the endothelial lining and subendothelial connective tissue with progressive stenosis of vascular lumen.[13] Radiation-induced biochemical alterations and stressogenic stimuli can drive morphological and functional alterations in endothelial cells, remodeling of tight junctions and neurovascular units interactions, increases in permeability of endothelial lining of microvasculature, cell death, and detachment from the basement membrane.[5]

The major aspect of RIMS is the total/partial absence of collateral circulation as compared with traditional Moyamoya disease.[6] Patients may present ischemic symptoms also due to slight decrease of cerebral blood flow (CBF), with consequent severe disability. Several surgical techniques are available for revascularization, including direct indirect, or combined bypass technique.[2] In our opinion, the EDAMS with MBHS results in good collateral revascularization, improved cerebral perfusion, and excellent short-term and long-term symptom control, with low perioperative risk. [7]

Although RT is the major risk factor for later cerebrovascular events, another mechanism that may contribute to the onset of cerebral ischemia is the decrease of CPP with compromised brain oxygenation caused by the raise of ICP.[4,10]

The CBF is regulated by the Monroe-Kellie doctrine[8] which state that space of the cranial cavity is fixed in volume, so an increase of one intracranial component (brain,
CBF, or CSF) must, therefore, result in the loss of another component in equal amounts: there is an inverse correlation between CBF and CSF. In patients with ventricular shunt for hydrocephalus, a malfunctioning of the device can lead to a rapid increase of ICP undermining the CBF and resulting in possible tissue ischemia.

Figure 1: Coronal TSE T2-weighted image (a) shows the residual tumor mass that enchases the terminal ICAs. The coronal MIP reconstruction of TOF MR Angiography (b) shows lack of signal in the terminal part of the left ICA due to severe stenosis and slight reduction of signal in the distal MCA branches. The left ICA angiograms on frontal (c) and lateral (d) projections confirm the severe stenosis of the terminal part of the ICA, proximal M1, and A1 segments; slight evidence of basal moyamoya network is also observed. No significant narrowing is observed on the right ICA frontal angiogram (e).

Figure 2: Coronal TSE T2-weighted (a) and axial b-1000 DWI images (b) in acute phase of shunt dysfunction show dilated ventricles (asterix in 2a) and acute ischemic changes on MCA and ACA cortical territories, with restricted diffusion, effaced sulci and scissures and cortical edema (arrowheads in 2a). Early postoperative images (c and d) show progressive improvement of cortical ischemic areas; new-onset deep ischemic lesions in basal ganglia are also observed (arrows).
CONCLUSION

Children receive RT for brain tumor, particularly if the circle of Willis region is involved, require close surveillance for the development of vasculopathy and consequent stroke. In our opinion, this surveillance must be even tighter if the patient has been treated with ventricular shunt for the possible synergistic interaction between the two causes on CBF and the risk of acute ischemic events. In particular, symptoms of shunt malfunction should be promptly recognized, and adequate treatment configured.

Statement of ethics

Written informed consent was obtained from the parents of the patient for publication of this case report and any accompanying images. All identifying information were stripped off. The manuscript was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.

Author contributions

Conception of the work: FV, PS; Acquisition of data for the work: FV, DC; Drafting the work: FV, PS; Revising it critically for important intellectual content: GM, MRS; Final approval of the version: GC.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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