CASE REPORT

Occult moyamoya disease causing fulminant infarction after septorhinoplasty

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Abstract

Moyamoya disease is an extremely rare cerebrovascular condition that predisposes affected patients to stroke in association with progressive stenosis of the intracranial internal carotid arteries and their proximal branches. To our knowledge, this is the first report of a lethal complication due to moyamoya disease after septorhinoplasty. A 25-year-old female Caucasian patient presented to our outpatient clinic with impaired nasal breathing for septorhinoplasty. Regrettably the patient died 6 days postoperatively due to progressive infarct series affecting all major cerebral vessels. Despite a thorough knowledge of possible local complications after septorhinoplasty, it is of utmost importance to consider rare general complications like moyamoya disease. Although cerebral infarctions are very rare in young people, it is crucial to identify and correctly interpret underlying typical symptoms.

Introduction

First described in 1957, moyamoya disease is an extremely rare cerebrovascular condition that predisposes affected patients to stroke in association with progressive stenosis of the intracranial internal carotid arteries and their proximal branches. Further, reduced blood flow in the major vessels of the circulation of the anterior brain leads to compensatory development of collateral vasculature that can lead to haemorrhage [1]. On angiography, the formation of these fragile vessels appears as “a puff of cigarette smoke”, named moyamoya in Japanese [2]. Besides the aforementioned typical radiographic findings and the prevalence to Asian origin, moyamoya can also be associated with conditions such as radiotherapy of the head and neck, dwarfism and others [1,3].

Here, we report a young female Caucasian patient, who underwent functional septorhinoplasty and afterwards suffered from a series of cerebral infarctions due to an occult underlying moyamoya disease.

Clinical report

A 25-year-old female Caucasian patient presented to our outpatient clinic with impaired nasal breathing due to nasal deviation caused by a deviated septum (Figures 1 and 2). The patient’s medical history was significant for left-sided otosclerosis, arterial hypertension, dwarfism, thoracic kyphoscoliosis, several allergies, for example, nickel and synthetic flavours, and headache 1 week prior to surgery. Besides lercanidipine 5 mg/day for 1 week prior to hospital admission, no further medication was recorded. Previous surgeries for cholecystectomy and correction of scoliosis under general anaesthesia were uneventful. About 6 hours after functional septorhinoplasty under uneventful general anaesthesia, the patient complained of dysphonia and nausea. For symptomatic treatment of the nausea, granisetron 3 mg diluted in 100 ml sodium chloride solution were administered, and the patient was presented to the otolaryngologist to exclude laryngeal violation during tracheal intubation in general anaesthesia as cause for dysphonia. One hour later the patient additionally suffered from left haemiplegia and a fixed right-view. Computed tomography of the brain revealed a large subacute right middle cerebral artery infarct and a small subacute infarct in the left posterior cerebral artery. Cerebral angiography and magnetic resonance angiography revealed bilateral stenosis of the internal carotid and middle cerebral arteries with a puffy collateral vascular network typical for moyamoya disease (Figure 3). The patient was transferred to the stroke unit and further on underwent parietal craniotomy of the right side for intracranial pressure release. Trans-oesophageal echocardiography revealed no persistent ovale foramen or other cardiac anomalies; further investigation also showed no further risk factors for cerebral infarctions, besides the mentioned moyamoya disease. Regrettably the patient died 6 days postoperatively due to progressive infarct series affecting all major cerebral vessels.
Discussion

Cerebral infarctions in young adults are extremely rare with an incidence rate at 11.3 cases per 100,000 people per year in the white population [4]. Of them, approximately 10 – 14% occur in young adults aged between 18 and 45 years and their incidence, risk factors and aetiology differ notably from those in older patients. Among others, congenital or acquired heart disease, haematological and metabolic disorders, substance abuse and vasculopathies have been identified as prothrombotic conditions [5]. Among vasculopathies, moyamoya is a very rare entity, which majorly affects Asians but rarely Caucasians [1]. Besides the other already-mentioned associated conditions, it is reported that patients with moyamoya disease can also suffer from headache, arterial hypertension and dwarfism [1,3] – afflictions that we also saw in our patient. One day prior to septorhinoplasty the patient’s blood pressure was 150/90 mmHg, the anaesthesiology protocol revealed a minimum blood pressure of 115/60 mmHg during general anaesthesia. One week prior to surgery the patient suffered from headaches and arterial hypertension, for which she received lercanidine 5 mg/day from her general practitioner. Previous studies have shown that patients with moyamoya disease have very limited haemodynamic reserves and that just a moderate decrease in cerebral blood perfusion, as seen during antihypertensive therapy or during general anaesthesia, may predispose and lead to cerebral infarctions [6,7]. In our case the initial symptoms of a cerebral stroke such as dysphonia and nausea have been misinterpreted as results of the tracheal intubation and general anaesthesia, which led to a delay in the correct diagnosis.

In case of a devastating complication such as cerebral infarction after an elective procedure in a young patient, quick diagnosis, decision taking and an interdisciplinary approach may be crucial for patient’s survival and an adequate management of the particular complication. Although complications after septorhinoplasty are reported, they mostly affect the surgical site [8]. To our knowledge, this is the first report of a lethal complication due to moyamoya disease after septorhinoplasty. The patient reported passed out 6 days after septorhinoplasty. We feel obliged to report this dreadful complication, so that our fellow surgeons may have the opportunity to correctly identify evidence as well as symptoms of underlying moyamoya disease and thus enable them avoid such disastrous complications in the future.

We conclude that despite a thorough knowledge of possible local complications after septorhinoplasty, it is of utmost importance to consider rare general complications like moyamoya disease, even more whenever a patient
suffers from associated afflictions such as headaches, arterial hypertension and dwarfism. Although cerebral infarctions are very rare in young people, it is crucial to identify and correctly interpret underlying typical symptoms.

**Declaration of interest:** The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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