

Bifid uvula: Anesthetist don't take it lightly!

Sir,

Bifid uvula means a cleft in uvula. It is often considered as a marker for sub mucous cleft palate. Compared to the

normal one, it has fewer amounts of muscular tissues. It is commonly noticed in infants and is rarely found in adult. It can cause problems in ear. Sometimes it is unable to reach the posterior pharyngeal wall during swallowing, causing

regurgitation. It may produce velopharyngeal insufficiency and nasal intonation.^[1] It does not cause many problems in view of airway management. It may be associated with aneurysm in different vascular bed like coronary and abdominal aortic aneurysm. We are presenting herein a case of bifid uvula with normal airway anatomy that was initially posted for retinal surgery, postoperatively diagnosed as cerebral aneurysm rupture, and later underwent a clipping surgery. To the best of our knowledge, there is no report on the association of cerebral aneurysm in bifid uvula patient with such a devastating complication in a 16-year-old boy. We obtained written consent from the parent of the patient to report this case.

A 16-year-old boy, weighing 52 kg, was presented in preanesthetic clinic for fitness for retinal surgery. He is of American Society of Anesthesiology physical status class I, with all other blood reports and chest X-ray within normal limits. Airway examination was normal, except for bifid uvula [Figure 1] and mild hypertelorism. No other clinical features were suggestive of any syndrome. Anesthetist cleared him for surgery. In view of aspiration, he was premedicated with ranitidine 150 mg and metoclopramide 10 mg at night and early morning respectively. He was induced with morphine 6 mg, propofol 120 mg, and vecuronium 6 mg. During intubation, 4 min after induction, his non-invasive systolic blood pressure increased to 210 mmHg due to laryngoscopic response. Immediate 20 mg propofol bolus was given in addition to induction dose. Intra-operative hemodynamics remained within acceptable range. He developed T-wave inversion in Lead II (may be juvenile T-inversion). Surgery took 180 min. We planned for full awake extubation and used esmolol 30 mg to prevent extubation response. The patient was not responding to command ($E_2V_1M_5$ -Glasgow Coma Scale; GCS) even after full muscle relaxant reversal (train-of-four ratio 97%). His bilateral pupils showed anisocoria. We waited for next 1 hour, but there was no change in the GCS. He was then shifted to intensive care unit (ICU), and computed tomography (CT) was performed after consulting the neurologist and neurosurgeon. CT showed ruptured aneurysm in the middle cerebral artery [Figure 2]. Immediate nimodipine was started, and radiological and cardiological evaluations were done. Then, the patient was shifted in neurosurgical theatre for clipping. Craniotomy and clipping surgery was done while maintaining a stable hemodynamics and shifted with intubated. Postoperative ICU evaluation failed to discover any other congenital anomalies. He was successfully extubated and discharged to home.

Bifid uvula, although looks benign, apparently sometimes may be associated with anomalies leading to catastrophic complications. Cornelia de Lange syndrome is a rare



Figure 1: A 16-year-old boy with bifid uvula

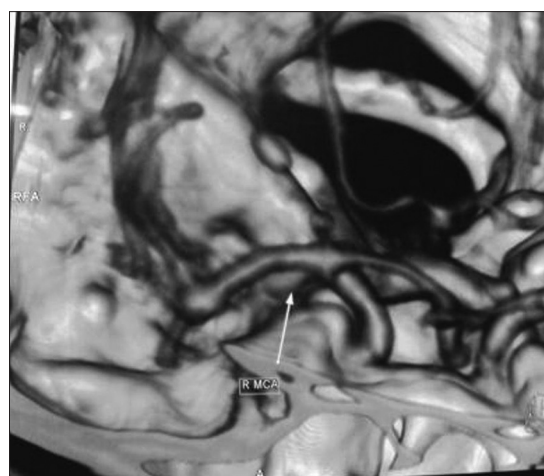


Figure 2: Ruptured aneurysm in the right middle cerebral artery of the patient

congenital syndrome associated with bifid uvula and sub mucous cleft palate that causes problems in airway due anatomical distortion.^[2] Bifid uvula may be associated with increased risk of schizophrenia, mild mental retardation, and chromosomal disorder, diagnosed by fluorescent *in situ* hybridization technique.^[3] Loeys–Dietz syndrome (autosomal dominant) is a genetic syndrome with clinical features overlapped with Marfan syndrome, but etiology due to mutations in the genes encoding transforming growth factor beta receptor 1. Hypertelorism, cleft palate, or bifid uvula are the major findings. Arterial aneurysms/dissections, arterial tortuosity involving aortic and its branches, carotid, vertebral, extracranial artery, abdominal aorta and its branches, common iliac, and popliteal arteries are reported in this syndrome.^[4,5] In our case, bifid uvula may have been a warning sign of the syndrome with internal anatomical or functional changes without any external manifestation akin to the tip of an iceberg. Although cerebral aneurysm is very rare with bifid uvula, it may be a part of the abovementioned syndromes. Thus, whenever anesthetists plans to conduct

a case with bifid uvula (even though non-syndromic), they must ask for detailed family and genetical history, clinical examination relevant investigations, and specialty consultation. Adequate preoperative preparation and, accordingly, intra-operative management can prevent unexpected complications in such patients.

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