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Fatal hemorrhage after tonsillectomy

A case of segmental mediolysis in arterioles

Introduction

Fatal hemorrhage after tonsillectomy remains a rare but important complication. It has been attributed to a variety of pathophysiological causes, among them congenital malformations, genetic defects, infectious diseases as well as technical reasons (■ **Tables 1**; [1–11]).

Segmental mediolytic arteriopathy (SMA) is a rare condition. It was first defined in 1976 by Slavin et al. [12] and has been well described in the literature, although to date the etiology of the condition is unknown. In most case reports, SMA is diagnosed retrospectively once tissue has undergone histological examination as an uncommon, nonatherosclerotic, noninflammatory arteriopathy. All publications available describe that it affects medium-sized splanchnic branches of the aorta along with renal, carotid, cerebral, and coronary arteries (■ **Table 2**; [13–25]).

This article reports an unusual case of fatal bleeding after a bilateral tonsillectomy that was at least partly caused by SMA affecting arterioles only in the neck region in a 20-year-old woman.

Case report

A 20-year-old woman with secondary bleeding following a tonsillectomy was admitted to the central emergency department while being resuscitated. According to the emergency physician she was initially still responsive but showed massive bleeding and shortness of breath. Upon arrival at the clinic, pulseless elec-

trical activity was visible in the electrocardiogram (ECG). Immediately after admission to the emergency room, transoral hemostasis was started because of diffuse bleeding from the right palatine tonsil area without a clearly visible source.

Numerous ligatures were visible in the right tonsil bed, which could be easily removed by suction. The tissue appeared to be vulnerable and fragile. A coniotomy was performed due to unsuccessful intubation. Within this framework, a me-

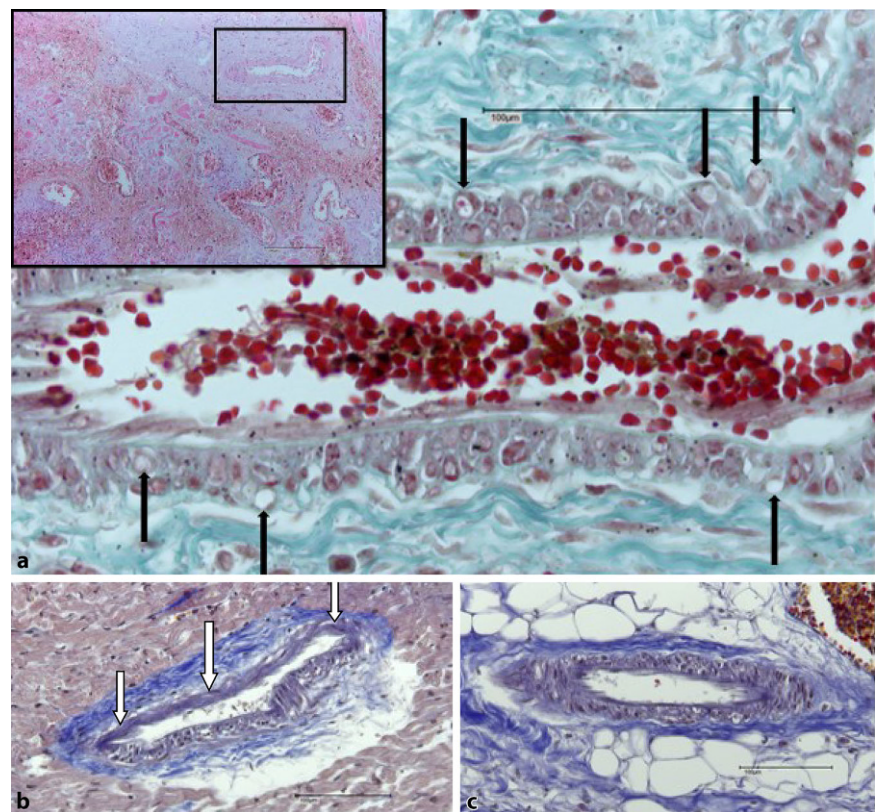


Fig. 1 ▲ **a** Peritonsillar area with arteriole (black box) about 2 cm distant from the original tonsillar resection margin (left lower corner), which is surrounded by diffuse bleeding not reaching the arteriole. (HE; bar 1000 μ m). Figure shows details of arteriole with mediolysis by vacuolization of medial muscle cells (black arrows), while the endothelial layer is intact; no thrombi or inflammation are present. Masson staining; bar 100 μ m. **b, c** Arterioles nestled in the myocardium. **b** Shows a segment with complete loss of nuclei of muscle cells of the media (white arrows), while the right (**c**) arteriole shows an intact vascular wall. SFOG; bar 100 μ m

Table 1 Causes of bleeding after tonsillectomy

| # | Cause of bleeding | References |
|----|---|--|
| 1 | Hyponatremia | Agut Fuster et al. [1] McRae et al. [2] |
| 2 | Congenital venous malformation | Foley et al. [3] |
| 3 | Emboli of pulmonary arteries | Green et al. [4] |
| 4 | Spontaneous | Griffies et al. [5] |
| 5 | Coblation technique | Heidemann et al. [6] |
| 6 | Infectious mononucleosis | Johnsen et al. [7] |
| 7 | Growth arrest-specific gene 6 | Laurance et al. [8] |
| 8 | Risk factors: age < 3 years; female, duration of surgery, incomplete hemostasis, coagulopathy | Sánchez Legaza et al. [9] |
| 9 | Aberrant carotid artery | Shihada et al. [10] |
| 10 | Electrosurgery | Windfuhr et al. [11] |

Table 2 Arterial mediolysis: locations, age/sex, associated conditions

| # | Location | Age (years)/sex | Associated conditions | References |
|----|---|-----------------|---|------------------------|
| 1 | Hepatic arteries | – | – | Armas et al. [13] |
| 2 | Hepatic and splenic arteries | – | – | Chan et al. [14] |
| 3 | Abdominal arteries | – | The middle colic artery, accounting for 14 (50%), followed by gastric and gastroepiploic arteries, (6 and 5, respectively) | Inada et al. [15] |
| 4 | Abdominal arteries | – | Post-transplantation lymphoproliferative disorder of the lungs | Khan et al. [16] |
| 5 | Mesenteric artery | – | – | Kato et al. [17] |
| 6 | Systemic, cerebral and pulmonary arteries | – | 5 cases of unsuspected SMA: 3 of visceral arteries; 1 case each affecting the cerebral and pulmonary parenchymal arteries | Lie [18] |
| 6 | Cerebral arteries | – | Stroke | Leu [19] |
| 7 | Pulmonary | 21 f | – | Müller et al. [20] |
| 8 | Internal carotid artery | 29 f | Right hemispheric infarction | Peters et al. [21] |
| 9 | Intracranial and intra-abdominal arteries | 48 m | Right vertebral, left internal carotid, superior mesenteric, bilateral renal; left external iliac | Sakata et al. [22] |
| 10 | Central, visceral and peripheral arteries | 27 m | Acute neurological defects involving his legs and urinary system: "string of beads" small aneurysms of intracerebral and extracerebral, renal, visceral and peripheral arteries | Schönefeld et al. [23] |
| 11 | Celiac to splenic and left renal arteries | 46 m | Simultaneous involvement of the left renal and right common iliac artery; adrenocortical adenoma, gastrointestinal stromal tumor, hepatocellular carcinoma and schizophrenia | Takagi et al. [24] |

SMA segmental mediolytic arteriopathy

chanical resuscitation was performed after a few minutes. Approximately 25 min later the decision to ligate the right external carotid artery was made because of the unstoppable diffuse bleeding. In addition, several blood transfusions had to be given. Because of a bleeding from the coniotomy site, the decision for a tracheostomy was made, which significantly improved the patient's ventilation. The preparation of the nerves and vessels on the right side of the neck was difficult during ongoing resuscitation. The patient's ventilation and coagulation deteriorated noticeably, so that after 1.5 h resuscitation measures were discontinued.

According to the enclosed medical records the woman had a tonsillectomy 10 days previously. 5 days before the fatal event she had an arterial secondary bleeding in the middle third of the left palatine which had to be clinically treated. There was no family history of bleeding or clotting disorders.

Autopsy findings

The autopsy was performed at the Institute of Forensic Medicine, University of Cologne, given that the primary question was one of medical malpractice. The autopsy showed a recent removal of both palatine tonsils. The surgical areas each had a diameter of about 5 cm, reached the base of the tongue and showed swollen and bled-in tissue. Inflammatory changes or fibrinous plaques were not visible. In the left palatine tonsillar bed, a surgical occlusion of a larger vessel was also detectable. The right palatine tonsil bed showed a clearly intense blackish bleeding. In order to identify a possible arterial source of bleeding, an extensive in situ preparation of the right tonsillar bed was performed, but no major vascular erosions, especially of the branches of the right lingual artery, were visible, not even later on the formalin-fixed preparation.

In addition, the autopsy showed signs of massive blood loss from the previously mentioned surgical areas, especially on the right side, as well as massive blood inhalation. During the autopsy blood was detected in the oral cavity, pharynx, epiglottis, esophagus as well as coagulated

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Fatal hemorrhage after tonsillectomy. A case of segmental mediolysis in arterioles

Abstract

This is the first report of a segmental mediolysis occurring in arterioles, affecting the neck/tonsillar region and leading to a fatal hemorrhage after a tonsillectomy in a 20-year-old woman. Arteriolar mediolysis could be observed in tissue layers about 2 cm distant from the tonsillectomy region. Reparative changes were observed in the carotid and small neck arteries as well as in only one intramyocardial arteriole; however, no other vascular regions typically reported in segmental mediolysis arteriopathy were affected and aneurysm formation was not observed. No family history of vascular disease or hypertension was known. Pathophysiologically, previously reported reasons for post-tonsillectomy bleeding could be excluded. Segmental mediolysis may represent a wider spectrum of affected arteriolar vessels besides the reported small and medium sized arteries leading to aneurysms, which impedes radiographic diagnosis.

Keywords

Arteriolar vessels · Vascular disease ·
 Posttonsillectomy bleeding · Neck region ·
 Reparative changes

in the stomach and the entire intestinal tract (small and large intestines). In addition, fresh liquid blood was found in the trachea and in the central airways. In the peripheral airways, liquid as well as coagulated blood were found. Incisions in the lung tissue revealed blood inhalation sites. Extensive intensive care measures revealed that the remaining lung tissue was significantly more fluid-rich and blood-rich.

At the same time, the emergency hemostasis measures on the right side of the neck were detectable during autopsy. A 10 cm long, fresh surgically treated skin incision was present in projection onto the anterior margin of the right sternocleidomastoid muscle. With further preparation, the vascular strand could be depicted in depth at the front edge of the right sternocleidomastoid muscle. The right external carotid artery was doubly ligated and severed in the middle. No vascular erosion could be detected with the naked eye in this area.

No further pathological changes of the internal organs were detectable, particularly no aneurysm formation or major bleeding source was found.

A genetic analysis of the vascular lesions was attempted (MVZ Martinsried, Germany); however, the autopsy material was already too degenerated following 3 years of storage.

Histological findings

The result of the pathological anatomical and microscopic examination of the surgically resected tonsils revealed a chronic and recurrent cryptitis and parenchymal tonsillitis on both sides. Vessels did not display visible loss of medial muscle cells or signs of destruction of the vessel wall (data not shown). Near the tonsillar resection regional fibrinoid necrosis and complete destruction of the vessel wall was observed, leading to extravascular bleeding. Occasionally, organized thrombi with revascularization were found [14]. The surgical resection margin showed regional bleeding into the tonsillar area with congested blood vessels. Differences could be observed between both resection areas: the left side showed signs of regeneration with multi-

ple capillaries and occasionally thrombi in organization in small arteries. The right tonsillectomy area revealed individual vessels with destruction of the vascular wall and extensive bleeding into the perivascular space. (■ Fig. 1a). Deeper tissue layers revealed arterioles with vacuolization of muscle cells in the media (■ Fig. 1; Masson staining). The endothelial layer was intact (Masson and CD31 immunohistochemistry, latter data not shown). The lesion showed neither inflammation nor signs of regeneration. One intramyocardial artery showed segmental loss of medial muscle cell nuclei in comparison to adjacent vessels (■ Fig. 1b, SFOG (Sour fuchsin orange G) and white arrows), while another myocardial arteriole (■ Fig. 1c) showed an intact vascular wall. In ■ Fig. 2 serial sections of an arteriole with segmental loss of muscular media are shown leading to segmental dilatation/narrowing along the course of the vascular lumen. Other arterial vessels of the neck, such as the carotid artery, displayed a segmental loss of the outer two thirds of the muscular media, with no tissue reaction such as inflammation visible, while the majority of the artery showed a regular medial wall (■ Fig. 3a). Another small artery displayed a similar morphology (■ Fig. 3b,c) with an almost complete transmural muscular loss in the media. Arterial vessels in other organs/organ systems such as the abdomen, kidneys, liver, coronary and cerebral arteries and all veins investigated did not reveal any aneurysm expected in mediolysis or similar pathology as described.

Discussion

Posttonsillectomy hemorrhage is a serious, albeit rare, and known complication after tonsillectomy, reported in 229 out of 15,218 adults (1.5%) [26] or less than 4% of tonsillectomized children. It was reported to have caused at least 4 deaths in a case series including 1,778,342 children [27]. It occurs predominantly early after surgery. Male patients (70 years of age or older), infectious mononucleosis, and a history of recurrent tonsillitis were identified as risk factors for post-tonsillectomy hemorrhage [26]. Delayed

hemorrhage has the potential to be life-threatening.

This report analyzed the histological changes leading to the death of a 20-year-old female patient after bilateral tonsillectomy with significantly delayed (left side 5 days and right side 10 days) postoperative secondary hemorrhage. Because no direct or indirect vascular injuries, postoperative inflammatory changes or vascular erosions were visible during autopsy, the question of pre-existing histopathological alterations/anomalies of the young patient's vessels came into focus with respect to the rare event of bilateral bleeding.

While the vascular lesions found were in accordance with those reported in

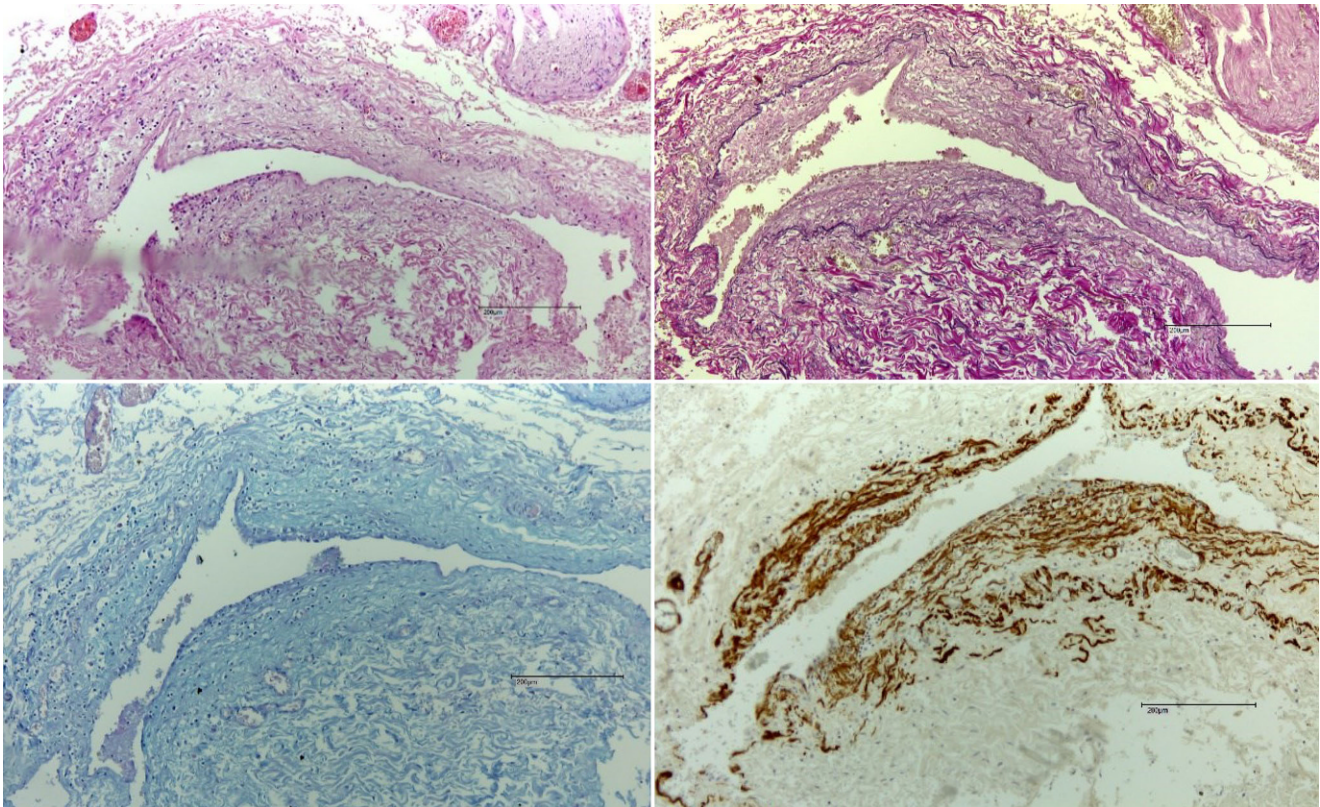


Fig. 2 ▲ Serial sections of arteriole (top: Hematoxylin-eosin stain (H&E); Elastika-van-Gieson stain (EvG); bottom: Arteriopathy (AP); segmental mediolytic arteriopathy (SMA)) with focal complete loss of the medial muscle layer (lower right, SMA immunohistochemistry), circumscribed pseudo-aneurysmatic dilatation and focal luminal narrowing. Each bar 100 µm

cases of segmental arteriolar mediolysis since their first detection in 1976 [12], the particular value of this report lies in the unique distribution of the lesions and the kind of vessels involved. In contrast to all reports available in the literature (Table 2), all relevant, i.e., death-related lesions, were exclusively found in the arterioles of the neck and tonsillar region. Spontaneous arterial dissection with pathognomonic signs of intimal tearing, mural hematoma or a double arterial lumen were not observed [28]. The nature of the injurious lesion was a vacuolar degeneration of smooth muscle cells as previously described [29, 30] but occurring in arterioles and not in larger arteries (Fig. 1a). Segmental lack of smooth muscle nuclei indicating loss of smooth muscle cells could also be observed in intramyocardial arterioles (Fig. 1b,c) and focally in arteries such as the carotid artery without causing aneurysms; however, no injurious lesions of small and medium arteries anywhere in other organs/organ systems were ob-

served during a full body autopsy. The formation of aneurysms with or without dissection of the vascular wall [21], potentially forming a “string of beads” pattern, were nowhere noted [22].

The assumed pathophysiology of SMA varies according to the literature. Fibromuscular dysplasia (FMD, [31, 32]) is a nonatherosclerotic, noninflammatory disease of the blood vessels that causes abnormal growth within the wall of an artery, which leads to hypertension at the mean age of 47 years. While segmental changes such as those observed in Fig. 2 may have superficial similarities to FMD, the observed variation in vessel wall thickness is obviously not due to additional growth but surviving muscle fibers while neighboring fibers have perished. Other diseases mimicking mediolysis such as connective tissue disorders (e.g. polyarteritis nodosa, Kawasaki disease, primary granulomatous central nervous system vasculitis, Wegener granulomatosis, Churg-Strauss syndrome, microscopic polyangiitis, Henoch-Schoen-

lein syndrome, systemic lupus erythematosus, rheumatoid vasculitis, and Behcet syndrome) [33, 34], Gsell-Erdheim medionecrosis [19] and infections (e.g., mycotic aneurysms [35]) have been discussed in the literature [36]. In our patient, we could exclude all of these potential mechanisms. No family history of SMA was recorded. Thus, this is the first case of a SMA occurring after tonsillectomy in arteriolar vessels.

Since there was a history of operations predating the occurrence of SMA, acute vasospasms due to inappropriate reactions to catecholamine or endothelial dysfunction [36] or endothelin-1 as surrogate vasoconstrictor [5, 37] might be considered. An arterial hypertension was not reported.

Presently, no genetic or familial components have been identified in any of the cases reviewed [25]. Since genetic testing on patients diagnosed with SMA may help identify potential familial genetic associations, we attempted to find genetic alterations potentially causing muscular

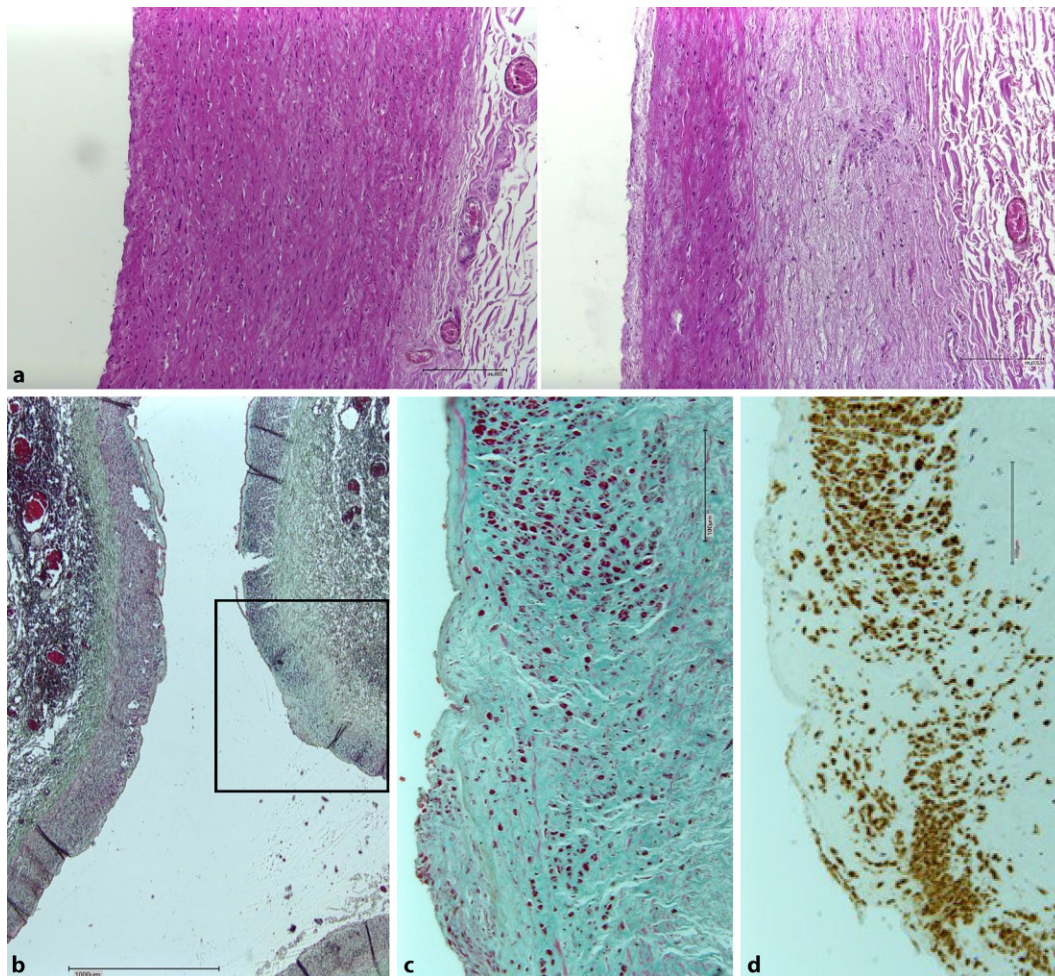


Fig. 3 ◀ **a** Carotid artery, different segments of the same vessel. Left: intact vascular wall; right: loss of outer and middle third of muscular layer of the media; no inflammation. HE; bar 200 μ m. **b** Low power longitudinal section of medium-sized artery depicting focal loss of media (black square). **c** Loss of smooth muscle, Masson stain, bar 100 μ m. **d** SMA immunohistochemistry with loss of smooth muscle throughout the media. Bar 100 μ m

mediolysis; however, the poor preservation of the autopsy tissue prevented further analysis.

In summary, we report the first case of a vascular mediolysis affecting arterioles predominantly in the tonsillar/neck region associated with bilateral tonsillectomy in a 20-year-old patient. While it enlarges our understanding of the spectrum of SMA, radiologic diagnosis is greatly impeded due to the vessel size. Thus, a mediolysis of arteriolar vessels may be considered as a potential cause of a diffuse bleeding after an operation (e.g. tonsillectomy) in a young patient. While we are not able to unequivocally prove a causal relationship, the coincidence of delayed bilateral bleeding and a vascular malformation merits in our opinion this case report as an extraordinary example of a routine operation gone wrong for a most unlikely reason.

Key points

- We report the first case of a vascular mediolysis affecting arterioles predominantly in the tonsillar/neck region associated with bilateral tonsillectomy.
- While fibrinoid necrosis and vascular bleeding was present in the immediate vicinity of the tonsillectomy region, arterioles in deeper tissue levels revealed vacuolization of medial muscle cells considered characteristic of a mediolytic pathophysiology.
- Mediolysis of arteriolar vessels may be considered as potential cause of a diffuse bleeding after an operation in a young patient.

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Declarations

Conflict of interest. K. Feld, W. Mellin, B. Melzer, M.A. Rothschild and J. Fries declare that they have no competing interests.

All procedures performed in studies involving human participants or on human tissue were in accordance with the ethical standards of the institutional and/or national research committee and with the 1975 Helsinki declaration and its later amendments or com-

parable ethical standards. The studies were performed in accordance with the guidelines of the central ethics committee and the German Medical Association.

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