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Vascular Malformation with Multiple Organized Thromboses in Buccal Region

Hiromitsu Yamamoto[1], Ken-ichiro Murakami[2], Mariko Kawai[1,3], Hitoshi Nagatsuka[4], Kazuhisa Bessho[1]

[1]Department of Oral and Maxillofacial Surgery, Graduate School of Medicine, Kyoto University, Kyoto, Japan
[2]Department of Oral and Maxillofacial Surgery, Ako City Hospital, Ako, Japan
[3]Department of Pharmacology, Osaka Dental University, Osaka, Japan
[4]Department of Oral Pathology and Medicine, Graduate School of Medicine and Dentistry, Okayama University, Okayama, Japan
Email: hyamamotokuhp@mac.com

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Abstract

Vascular malformations occur most frequently in the maxillofacial and oral regions. Although many cases of vascular malformations with phlebolithiasis have been reported, only few reports have documented cases of thrombosis formation. We report a rare case of a vascular malformation with multiple thromboses in the left buccal region. A 28-year-old woman had a painless swelling in the left cheek when she consulted our hospital. T1-weighted magnetic resonance imaging revealed a low-signal lesion, having the same intensity as the muscle, in the left buccal region, and T2-weighted imaging revealed a high-signal lesion. Additionally, three tuberous tumours were noted in this lesion showing both a light high signal on T1-weighted imaging and a low signal on T2-weighted imaging. Based on these imaging findings, as well as clinical and pathological findings, the patient was diagnosed with multiple vascular malformations in the left midfacial region. The three tuberous tumours showed necrotic tissue in the central area, which was formed by calcified and concentric fibrous tissue, and vascularization. Because tumorous lesion and typical vascular structure by the Elastica Van Gieson staining were not observed, these tumours were seemed that fibrin thrombus in this lesion formed the organized tissue.

Keywords

Vascular Malformation, Thrombosis, Tuberous Tumour, Buccal Region, EVG Staining

1. Introduction

Vascular anomalies in the soft tissue come under two categories: haemangioma/

haemangiogenic tumour and vascular malformation. Because they show multiple clinical findings, distinguishing between haemangioma/haemangiogenic tumour, which is an original tumour lesion, and vascular malformation, which is a congenital disease, is difficult. Therefore, these conditions are known by different disease and syndrome names. Mulliken and Glowacki advocated that these are completely different diseases on the basis of the characteristics of vascular endothelial cells [1]. Many researchers presently follow this concept [2].

Haemangioma is usually noted at birth, and grows proportionately as the child ages. It consists of abnormal, often combined, capillary, arterial, venous, and lymphatic vascular elements [3] [4]. In contrast, a vascular malformation is caused by abnormal vasculogenesis, appears at birth, increases gradually with age, and never disappears throughout life [5] [6] [7].

Although vascular malformations can develop in any part of the human body, they frequently develop in the head and maxillofacial region [5] [6] [7], and occasionally include phlebolithiasis [8] [9] [10]. Various vascular malformations have been reported to form thromboses [11]-[18], but few reports have documented their existence in the maxillofacial region [19] [20] [21].

We report a rare case of a 28-year-old woman who had a midfacial vascular malformation with multiple organized thromboses in the left buccal region, and document the treatment of this case with reference to the literature.

2. Case Report

A 28-year-old woman presented to the Department of Oral and Maxillofacial Surgery, Kyoto University Hospital, with a painless swelling in the left buccal region. About five years earlier, she had noticed a small painless swelling in the same region, for which she had sought treatment at another dental hospital. Magnetic resonance imaging (MRI) revealed a tumour in this region, and it was clinically diagnosed as a benign tumour. Although the swelling continued to grow gradually, only the follow-up was performed. Therefore, the patient consulted our department.

The patient's general condition and results of laboratory examinations were normal. The left side of the face was apparently swollen, with normal skin colour and no pain (**Figure 1(A)**). Three elastic, hard, movable, and spherical painless tumours could be palpated in the left buccal mucosa. They were approximately 30 mm, 20 mm, and 10 mm in diameter (**Figure 1(B)**). Moreover, two elastic, hard, and dark-violet painless tumours, approximately 18 × 10 mm and 15 × 8 mm in size, could be observed on the left tongue margin, and one same characteristic tumour, 5 × 5 mm in size, could be observed on the frenulum of the upper lip. T1-weighted MRI revealed a low-signal lesion, with the same intensity as the muscle, in the subcutaneous tissue of the left buccal region, extending from the frontal to medial region of the zygomatic bone (**Figure 2(A)**). This lesion also showed a high signal on T2-weighted imaging, suggesting the existence of a haematoma inside a benign tumour (**Figure 2(B)**, **Figure 2(C)**). Three tuberous

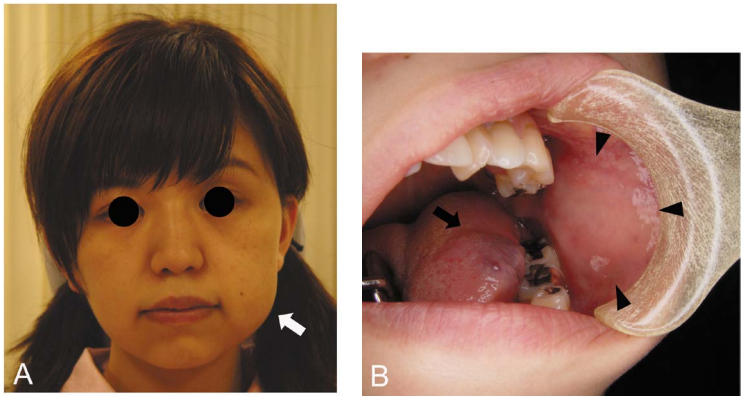

Figure 1. (A) The left cheek region of the patient was apparently swollen, with normal skin colour and no pain; (B) Three elastic hard, movable and spherical painless tumours could be felt in the left buccal mucosa. An elastic hard and dark violet painless tumours, approximately 18 × 10 mm size, could be also observed on the left tongue margin.

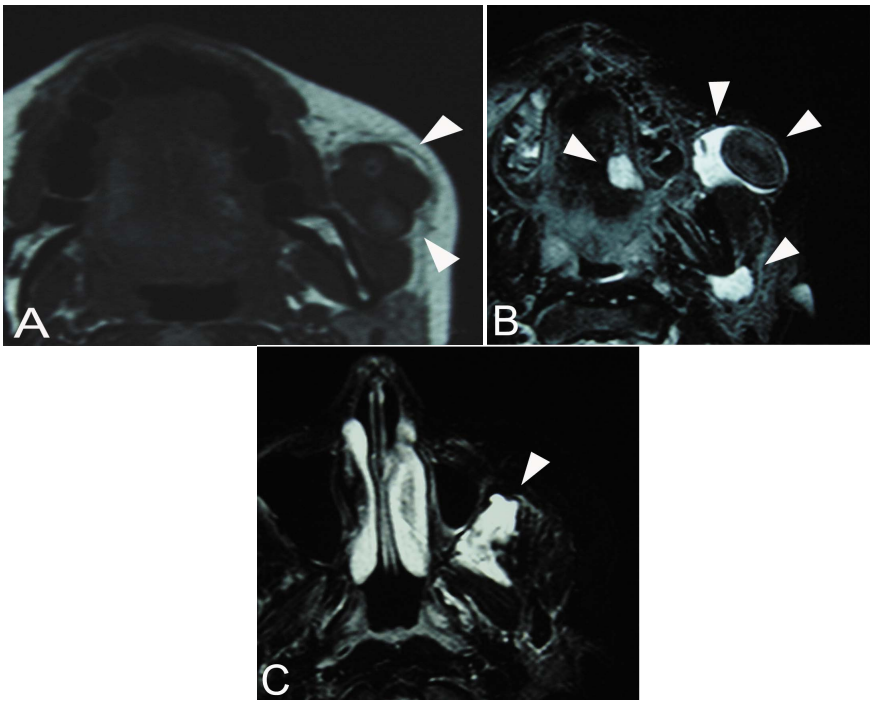

Figure 2. (A) A low-signal lesion, same intensity of the muscle, was observed in the subcutaneous tissue of the left buccal region on the T1-weighted imaging and spread from frontal to medial region of the zygomatic bone. (B) The lesion was shown a high signal in same region on the T2-weighted imaging and a tuberous tumour was shown in this lesion, which had segmented margin and were both a light high signal on the T1 and a low signal on the T2. Several lesions showing a high-signal imaging on the T2 same as the left buccal region were observed on the left tongue margin and in the left parotis. (C) Several lesions showing a high-signal imaging on the T2 were also observed in the area between the left orbita and zygomatic bone.

tumours were seen in this lesion, which had segmented margins and showed both a light high signal on T1-weighted imaging and a low signal on T2-weighted imaging (**Figure 2(A)**, **Figure 2(B)**). Furthermore, many lesions showing a high

signal on T2-weighted imaging, similar to those in the left buccal region, were observed in two areas in the left tongue margin, one area in the left parotid gland, and one area between the left orbital and zygomatic bones (**Figure 2(B)**, **Figure 2(C)**, and data not shown). These clinical and imaging findings suggested a diagnosis of multiple vascular malformations from the left buccal to midfacial region.

Based on the clinical and imaging findings, we considered this lesion a vascular malformation. Although there was no indication for a whole-tumour resection, we performed extirpation only of the three tuberous tumours in the left buccal region in order to improve the patient's cosmetic appearance. An approximately 20 mm incision was made on the left buccal mucosa under the parotid opening. Then, an elastic hard and white tumour, 15 mm in diameter, appeared after a search bluntly. It could be easily resected as it was not adhered to the surrounding tissues. However, severe pulsating bleeding occurred after tumour removal, and hence, astriction and haemostasis were performed using a vascular knot. An elastic, hard, and dark-violet tumour, 10 mm in diameter, was observed under this lesion. This tumour was also easily removed and resulted in similar bleeding, which was addressed as mentioned above. Moreover, under these lesions, another elastic, hard, and dark-violet tumour, 20 × 15 mm size, was observed. This again was removed and treated similarly to control bleeding. All tuberous tumours had similar characteristics, and were spherical and solid (**Figure 3(A)**, **Figure 3(B)**).

Histopathological analysis revealed that these three tumours had necrotic tissue in the central area, which comprised organized tissue formed by calcified and concentric fibrous tissue, and vascularization. Because tumorous lesion and the typical blood vessel structure by the EVG staining were not observed, these tuberous tumours were seemed that fibrin thrombus in this lesion formed the organized tissue (**Figures 3(A)-(G)**). The patient was discharged after satisfactory wound healing, and the facial views showed a fairly improved external appearance (**Figure 4**). Over five years, a recurrent tumour formation is not seen.

3. Discussion

Because vascular anomalies in the soft tissue, haemangioma/haemangiogenic tumour and vascular malformation, present various clinical findings, they are confusing to diagnose. In 1982, Mulliken and Glowacki suggested that they were different diseases based on the characteristics of vascular endothelial cells [1]. The International Society for the Study of Vascular Anomalies workshop also classified vascular anomalies [2].

Infantile haemangioma, a representative haemangioma, occurs within the first month of life, exhibits rapid proliferation, and slowly reduces to near-complete resolution [3] [4]. In contrast, a vascular malformation is a result of abnormal vasculogenesis, appears at birth, increases gradually with age, and never disappears throughout life [5] [6] [7].

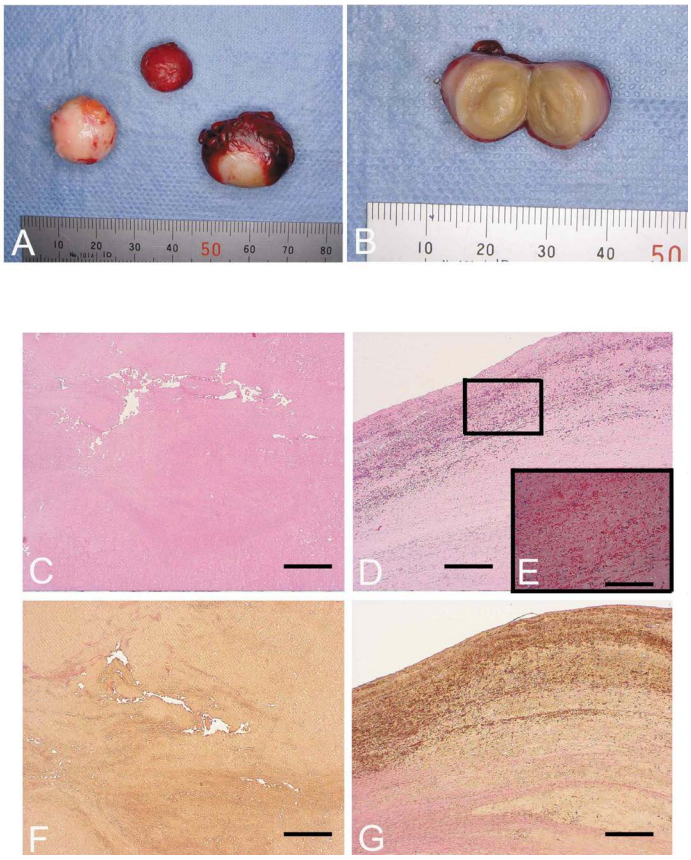

Figure 3. ((A), (B)) Extirpated three tuberous tumours have similar character, spherical and solid; (C)-(E) Histopathological findings in these lesions. These three tumours showed necrotic tissue in their central area, and they were the organized tissue formed by calcified and concentric fibrous tissue, and vascularization. Haematoxylin and eosin; ((C), (D)) original magnification, ×40; scale bars = 250 μm; (E) original magnification, ×200; scale bars = 50 μm. ((F), (G)): It was not observed that tumorous lesion and the typical blood vessel structure by the EVG (Elastica Van Gieson) staining. Original magnification, ×40.

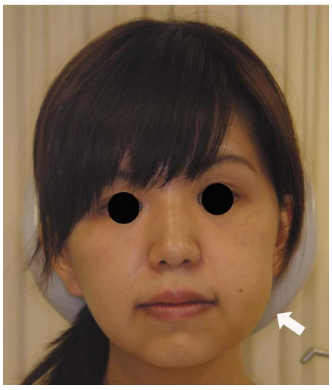

Figure 4. Facial views of the patient showed a fairly improved external appearance.

Vascular malformations frequently occur in the lip, tongue, and maxillofacial and oral regions [5] [6] [7]. Although several studies have reported vascular malformations occurring in the masseter muscle and buccal pad in the buccal or

midfacial region [22] [23] [24] [25] [26], or with phlebolithiasis [8] [9] [10], no previous report has documented a case of multiple organized thromboses. Despite not being able to confirm the final diagnosis, the current case seemed consistent with multiple vascular malformations in the midfacial region because of the following reasons: MRI and clinical findings that showed multiple dark-violet lesions like vascular malformations in the oral mucosa, clinical progress of this symptom from five years earlier, operative findings, and histopathological findings of the tuberous tumours [27]. Treatments for vascular malformations are simple surgical resection, cryosurgery, packaging treatment, and laser-assisted excision. In the absence of symptoms, several malformations are only followed up and not operated on, unless they pose a risk of complications or affect the patient's cosmetic appearance. In the present case, follow-up was selected at the first hospital because of the lack of apparent symptoms except the small left buccal swelling. However, since multiple tuberous tumours had appeared and the buccal swelling continued to increase gradually, which adversely affected the patient's external appearance, we planned to remove only these tumours for cosmetic purposes and for arriving at a histopathological diagnosis simultaneously. When organized thromboses in a vascular malformation are needed to extirpate, it is necessary to pay sufficient attention to bleeding as this case.

Three tuberous tumours were extirpated, and all had similar characteristics; they were elastic, hard, and spherical but of varying sizes. They did not show adhesion to surrounding tissues and were suspected to be some tumour of the salivary gland, such as pleomorphic adenoma. Histopathologically, all tumours showed necrotic tissue in the central area, which was considered organized tissue formed by calcified and concentric fibrous tissue, and vascularization. Internal fibrin thrombus seemed to have formed the organized tissue. Furthermore, we performed EVG staining, which specifically stains elastic and collagen fibres. However, all staining tests yielded negative results and did not reveal typical vascular structure. Although many such cases show partial growth of granulation tissue, the present case did not. Only a large necrotic area was shown. Such histopathological findings are extremely rare, since the patient was young and the tumours were multiple and large.

Although several reports have documented vascular malformations with thromboses in the central nervous system, heart, liver, kidney, pancreas, lumbar region, and eyes [11]-[18], few reports have documented them in the maxillofacial and oral regions [19] [20] [21]. Moreover, the mechanism underlying thrombosis formation in such vascular malformations has not yet been elucidated. Many vascular thrombo-haemorrhagic disorders present abnormal vascular findings, and thrombosis formation in such cases is closely related to blood effusion or haemorrhage. Generally, haemorrhage occurs because of many reasons, but they usually coagulate and finally form thromboses [28]. However, these disorders commonly occur as systemic syndromes, and hence, our case is different from them because of the differences in the patient's general condition and clinical findings.

Because of the lack of previous data, the possibility of recurrence of thromboses cannot be accurately predicted. However, since the main lesion remains in the midfacial region, thromboses may recur in the same area or other areas with multiple lesions. Therefore, periodic follow-up is necessary.

4. Conclusion

We report a rare case of a 28-year-old woman who had a midfacial vascular malformation with multiple organized thromboses in the left buccal region, and report the detailed treatment with reference to the literature.

Consent

Written informed consent was obtained from the patient for publication of this case report ant any accompanying figures.

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