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Case Report

Surgical excision of large scalp hemangiomatosis: a case report and literature review

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Abstract: Scalp hemangiomatosis, particularly large size scalp hemangiomatosis, are very rare and difficult to treat. The present report presents one case of large scalp hemangiomatosis. The patient is a 37 year-old female. The mass on the left parietal region of the head had existed for 14 years, gradually increased in area, and exhibited pulsation. Arteriovenous malformation was considered before surgery. After ultrasound and digital subtraction angiography (DSA) evaluation, the main feeding arteries were ligated. Sutures were placed around the mass to tightly clamp and control the blood supply. The mass was found to be rich in adipose tissue and blood vessels during surgery, and a pathological examination confirmed the presence of a hemangiomatosis. Postoperative scalp healing was poor, and scalp defects occurred. Local dressing change and iodoform gauze packing in the defect area resulted in complete scalp healing. The present case demonstrates that the scalp can exhibit large-size hemangiomatosis, which can be easily misdiagnosed as vascular malformations. Sufficient preoperative evaluations, including necessary examinations by ultrasound and DSA, are required to treat this disease. The feeding arteries should be clearly identified and ligated; suturing the tissues around the lesion is also effective for controlling blood loss during surgery. If possible, the lesions should be completely excised to expose the healthy scalp. If scalp defects occur, Local dressing change and iodoform strip packing can effectively promote scalp healing.

Keywords: Scalp, hemangiomatosis, surgical excision

Introduction

As a diffuse and continuous angiogenic disease, hemangiomatosis normally occur in subcutaneous tissue, muscle, bone, and adipose tissue. This disease typically involves a large area with a continuous distribution and is sometimes multifocal. Hemangiomatosis tend to frequently occur on the trunk and limbs [1]. Occurrence on the face and neck has also been occasionally reported [2]. However, the occurrence of scalp hemangiomatosis is very rare. A literature search produced only one case reported by Kayaselcuk et al. in 2002. In that report, due to the small size of the tumor, treatment was not difficult [3]. No cases of large scalp hemangiomatosis have been reported to date. The present paper reports an uncommon case of a large scalp hemangiomatosis. The blood was supplied from branches of the external carotid artery. Although it was difficult to treat, satisfactory surgical results were finally obtained. Due to its rareness, this case is presented here to report the treatment experience for the surgical excision of large scalp hemangiomatosis.

Case report

A 37 year-old female patient was hospitalized due to "progressive growth of a mass on the left parietal region of the head for 14 years and surface ulceration with scar tissue for 1 week". She was healthy prior to hospitalization. The mass on the left parietal region of the head, which was found 14 years prior, was small and soft and could be reduced upon compression. The mass had not been treated and had gradually grown in recent years. In the preceding week, the surface of the mass had ulcerated and a scar formed. Physical examination was performed. No positive symptoms were found in the nervous system. A 15 cm ×10 cm ×1.5 cm mass with a rough and uneven surface was found on left parietal region of the head. The scalp was filled with blood and exhibited red-

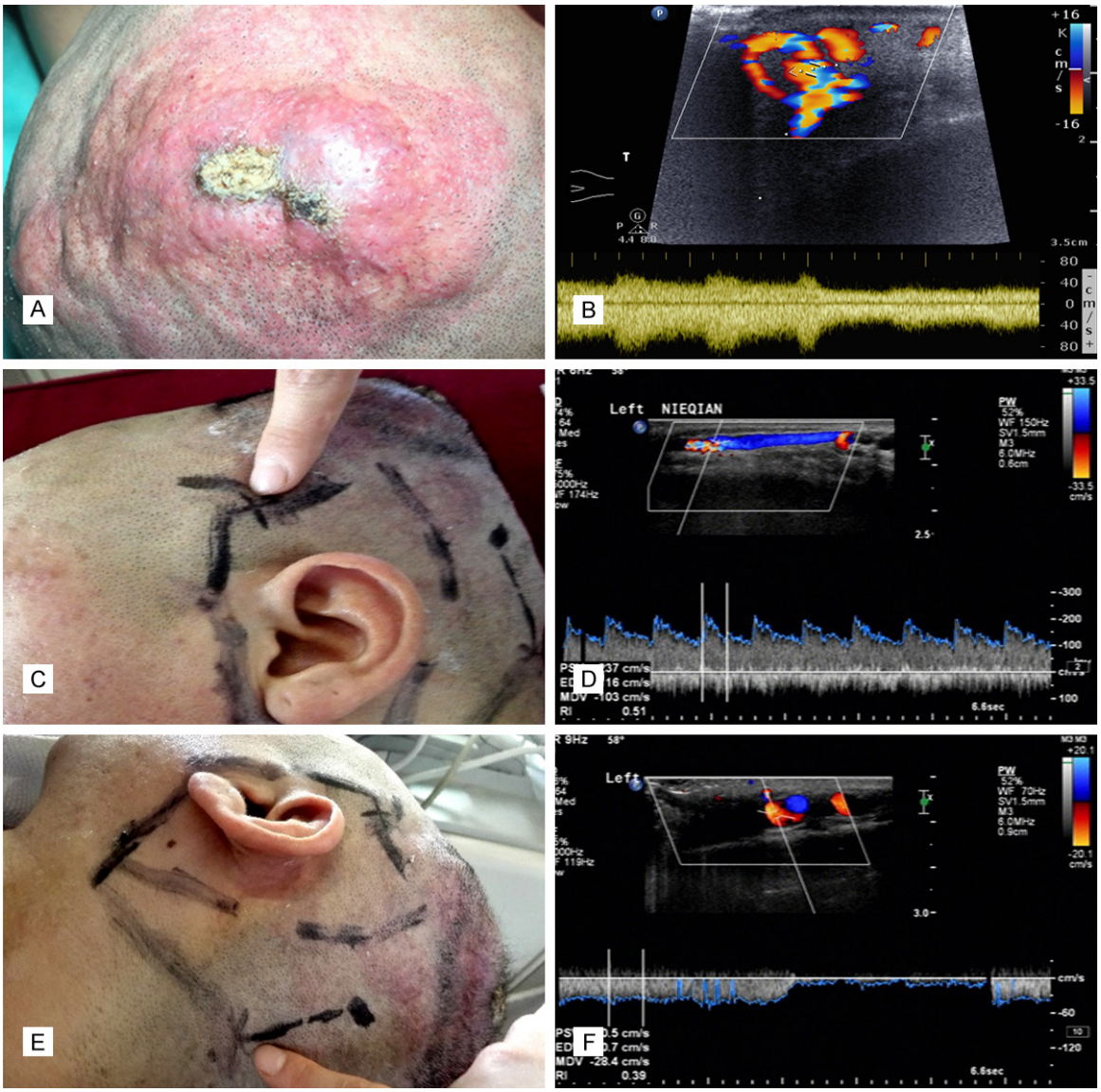

Figure 1. Ultrasound of the head. A. A scalp mass with bulging, redness on the surface, and forming a scar after ulceration in the center is shown on the left parietal region of the head. B. An ultrasound examination of the mass shows the tortuously expanded abnormal blood vessel structures with rich blood flow. C, D. The superficial temporal artery (indicated by the finger) was the main source of the blood supply; the diameter of the artery was 2.6 mm; the blood velocity was 237/116 cm/s; eddy flow was observed, and the pulsatility index (PI) was decreased. E, F. The occipital artery (indicated by the finger) was the secondary source of blood supply; the diameter of the artery was 2.1 mm; the blood velocity was 39.5/24.5 cm/s; eddy flow was observed, and the PI was decreased. The positions indicated by fingers are the primary pressure points of the superficial temporal artery and occipital artery. Pressurizing the left superficial temporal artery markedly decreased the blood flow to the mass, while pressurizing the left occipital artery slightly decreased the blood flow.

ness. Pulsation and vibration could be felt by touching the mass. The center of the mass was reddened, ulcerated. Expanding arteries and veins could be observed surrounding the mass. Pulsating and throbbing noises could be heard by auscultation. Color ultrasound of the blood vessels of the mass revealed large blood vessel-rich mass inside the scalp on the left parietal region of the head. Within the mass, a tortuously expanded abnormal blood vessel structure could be observed. The main sources of the blood supply were the left superficial temporal artery and the occipital artery. The blood flow to the mass was markedly reduced if the

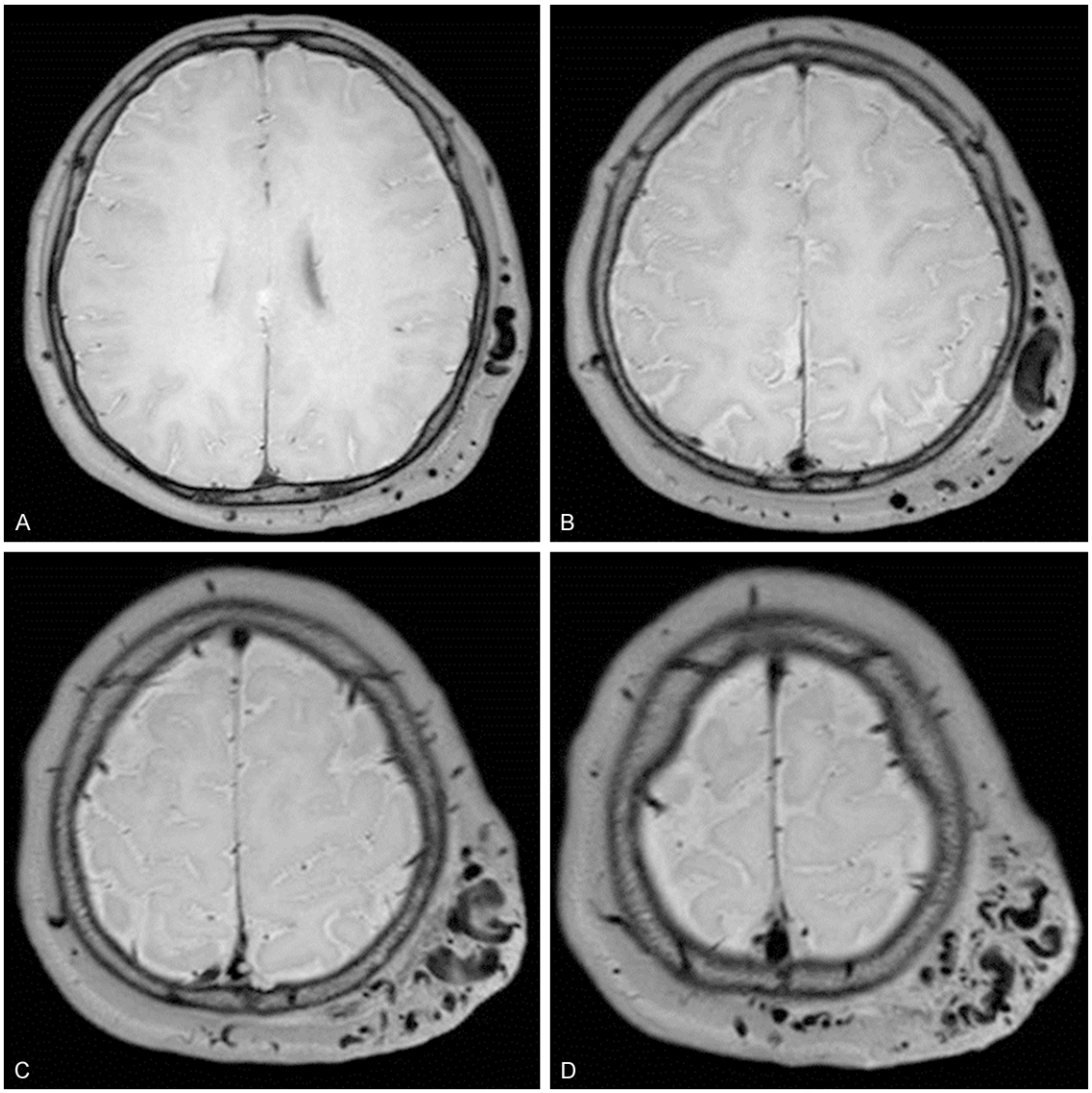

Figure 2. MRI examination of the lesion on the head. A-D. Different MRI sections revealed the blood vessel-rich mass inside of the scalp on the left parietal region of the head. Blood flow voids can be observed inside of the mass. The dimension of the lumen varied. The dimensions of the blood vessels of the surrounding normal scalp were increased. The lesion was localized to the scalp and had not invaded the skull.

left superficial temporal artery and occipital artery were compressed (**Figure 1**). An evaluation of the head using magnetic resonance imaging (MRI) showed large blood vessel-rich mass inside of the scalp on the left parietal region of the head. Blood flow voids could be observed inside of the mass. The dimensions of the lumen varied. The dimensions of the blood vessels of the surrounding normal scalp were increased. The lesion was localized to the scalp and had not invaded the skull (**Figure 2**). Further examination of the head by digital subtraction angiography (DSA) showed that the main sources of the blood supply to the mass were the left superficial temporal artery and the occipital artery; the right superficial temporal artery and occipital artery also passed over the mass and acted as secondary blood supply arteries. The blood flow of the mass primarily drained from the superficial temporal artery to the external jugular vein. Meanwhile, imaging of both sides of the internal carotid arteries and the vertebral artery revealed no blood supply to the mass from the intracranial

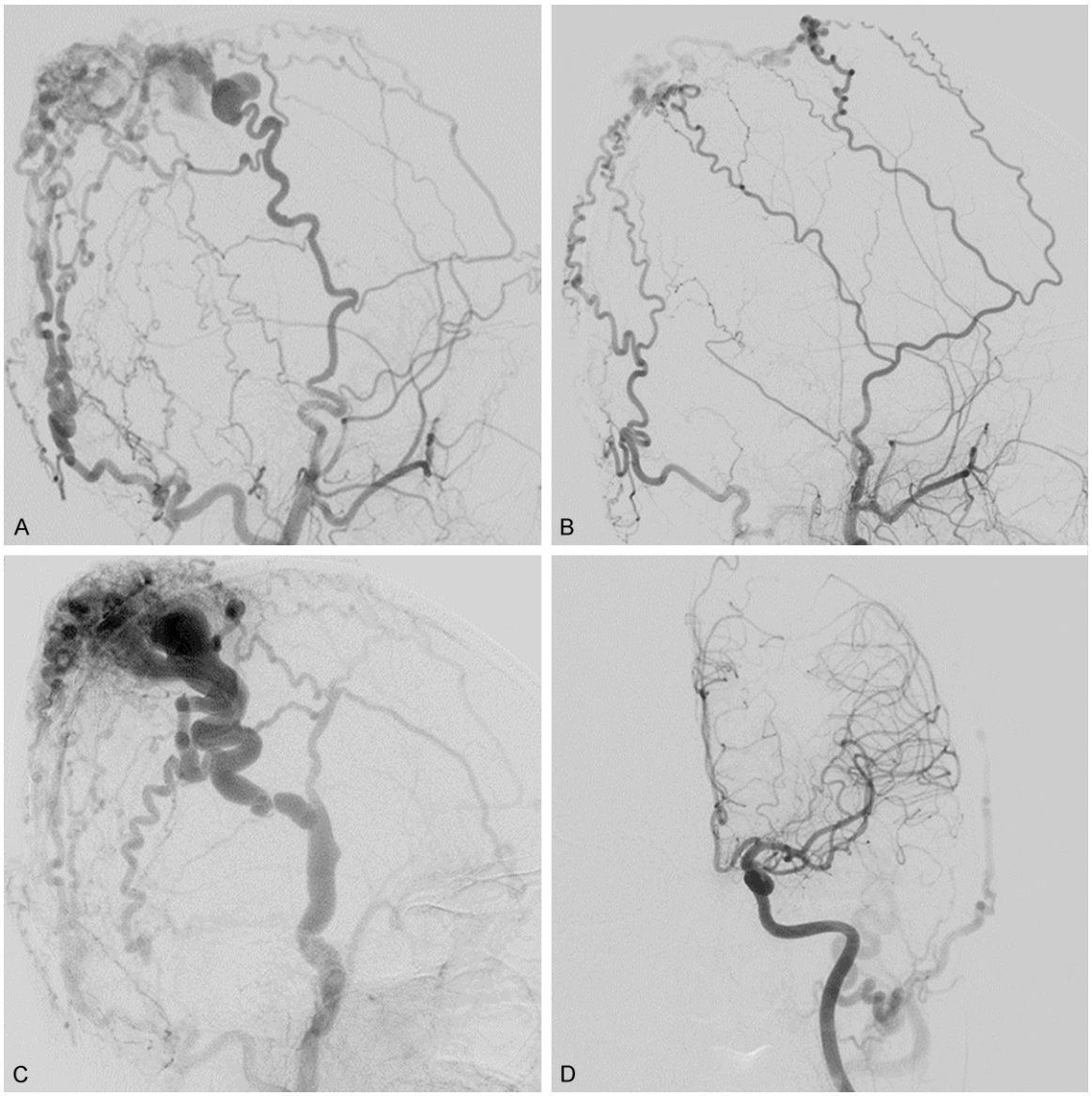

Figure 3. DSA examination of the head. A. Fluoroscopic imaging of the left external carotid artery revealed that the main blood supply sources were the superficial temporal artery and occipital artery. Artery expansion can be observed in the lesions close to the superficial temporal artery. B. Fluoroscopic imaging of the right external carotid artery revealed that the branches of the superficial temporal artery and occipital artery also contributed to the blood supply. C. Blood was drained to the temporal superficial vein from the lesion vein. D. Fluoroscopic imaging of the left external carotid artery shows that no veins in the skull are involved in the blood supply.

artery (**Figure 3**). Taking into account the clinical and medical imaging characteristics, the mass was diagnosed as an arteriovenous malformation, and surgical excision was planned.

The surgical incision shape was a shuttle cut with the lesion at the center. After labeling the superficial temporal and occipital arteries, sutures were placed around the mass to reduce the blood supply to the mass. The scalp was cut to expose the left superficial temporal artery and occipital artery for ligation. Then, an incision was performed along the surrounding area of the mass. A rich blood supply was observed in the mass during surgery. Using monopolar electrodes and bipolar coagulation, the lesion was excised, revealing rich adipose tissue and thickened blood vessels (**Figure 4**). After excision of the mass, the normal scalp surrounding the lesion was stretched; then, the

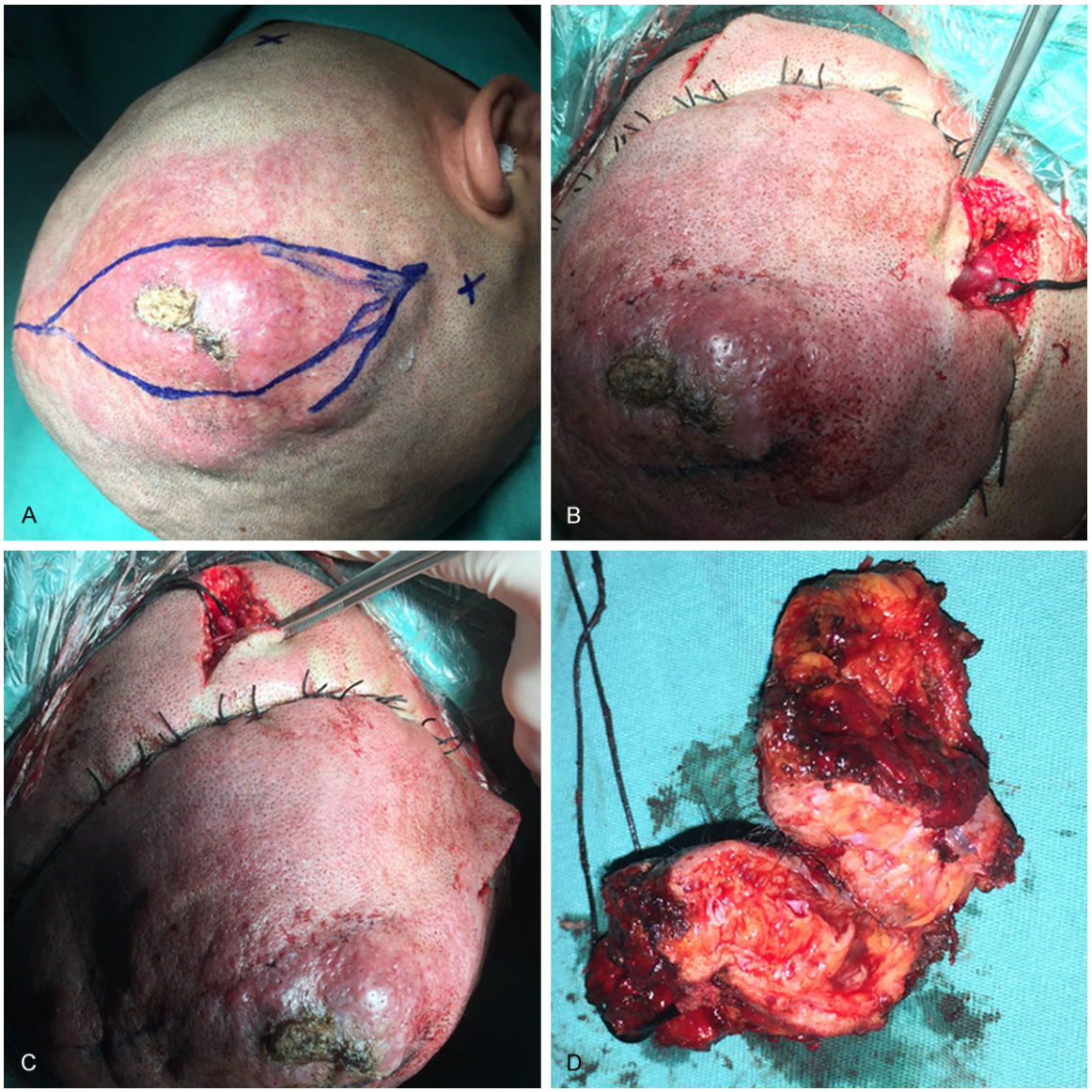

Figure 4. Photograph during surgery. A. The design of the surgical incision. ×-shaped labels correspond to the superficial temporal artery and occipital artery. B. Sutures were placed around the lesion to reduce the blood supply to the mass. The scalp was cut to expose the superficial temporal artery for ligation. C. The occipital artery was exposed and ligated. D. After incision, the lesion was opened to reveal adipose tissue and thickened blood vessels.

incision was sutured. After surgery, the mass was pathologically examined. The results showed that the lesion was beneath the epidermis and exhibited a transformation to a capillary hemangioma; expansion of small veins was observed. Venous, cavernous, and capillary blood vessels were mixed and randomly distributed in the fibrous adipose tissue beneath the epidermis. The walls of venous blood vessels were irregular, and the thinned lumen was expanded. The transformation to a capillary hemangioma was exhibited in part of the matured adipose tissue. Congestion was found in part of the lumen, and chronic inflammatory cell infiltration was found to be interstitially dispersed. No bacterial infection was detected in the tissues. Immunohistochemical staining was positive for CD31 and CD34 (**Figure 5**). The lesion was diagnosed as a hemangiomatosis from the pathological perspective. After surgery, the scalp was well aligned. However, after the removal of sutures, the scalp in the surgical area gradually became necrotic and exhibited poor healing, resulting in

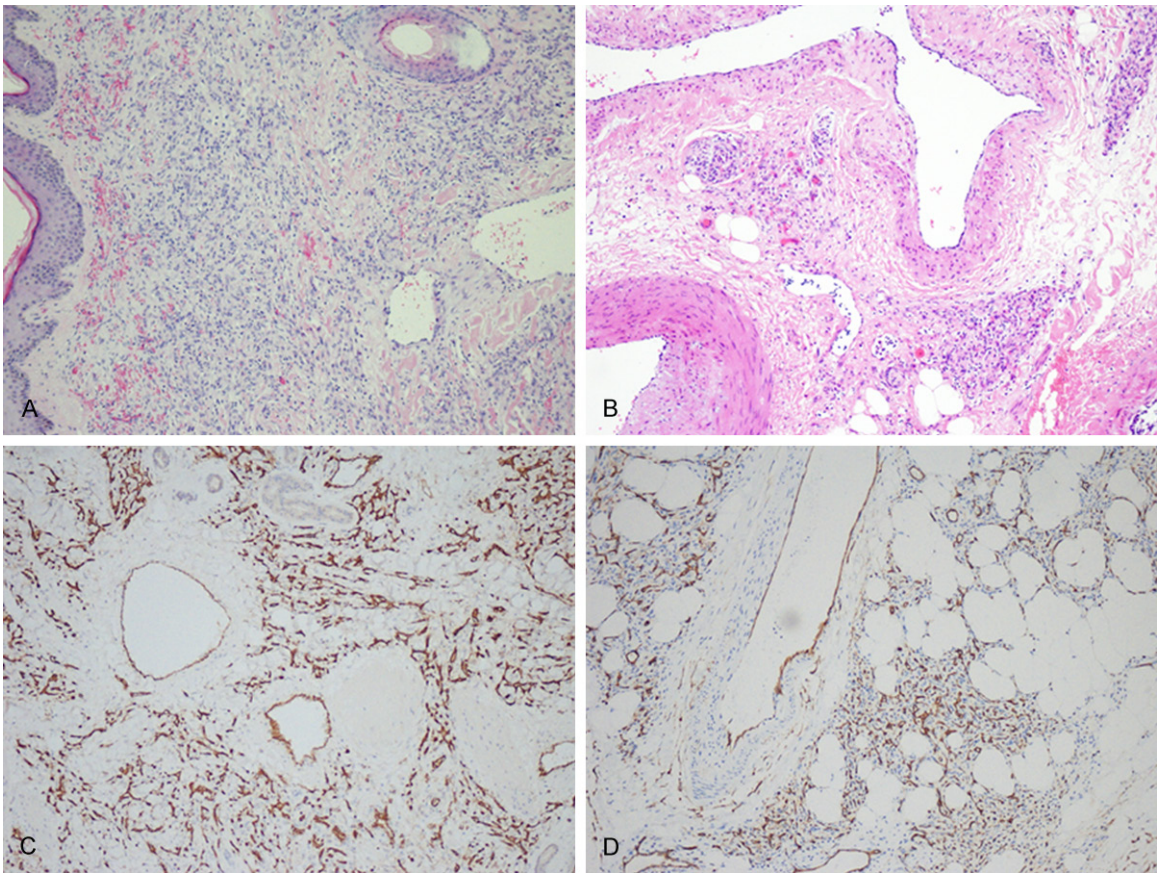

Figure 5. Pathological examination of the mass. A. (HE×100) The lesion was beneath the epidermis and exhibited transformation to a capillary hemangioma; the expansion of small veins was observed. B. (HE×200) Venous, cavernous, and capillary blood vessels were mixed and randomly distributed in the fibrous adipose tissue beneath the epidermis. The walls of the venous blood vessels were irregular, and the thinned lumen was expanded. The transformation to a capillary hemangioma was observed in part of the matured adipose tissues. Congestion was observed in part of the lumen, and chronic inflammatory cell infiltration was found to be interstitially dispersed. C. ×200, CD31 (+) staining. D. ×200, CD34 (+) staining.

defects in the scalp. Local dressing changes for the scalp defect and iodoform gauze packing were applied. After one month, a large portion of the scalp was growing well, and only the incision line exhibited poor healing. The scalp was completely healed within two months after surgery (**Figure 6**). MRI examination at 20 days after surgery showed that the lesion was completely excised and that the lumen of the expanding arteries in the surrounding tissue of the lesion had returned to normal (**Figure 7**). Postoperative follow-up over half of a year showed no sign of relapse.

Discussion

Hemangiomatosis is a benign disease primarily consisting of growing blood vessels and often accompanied by mature adipose tissue, fibrous tissue, lymph vessels, and nervous tissue and mainly occur on the trunk and limbs [1]. Although a hemangiomatosis a benign disease, it tends to invade and grow slowly and extensively or grow vertically into very deep tissue. Hemangiomatosis exert effects on skin, muscle, and bone tissue. These biological characteristics make surgical excision difficult [4]. Almost all hemangiomatosis occur in young females. The disease could be caused by congenital or acquired conditions. Congenital conditions include Klippel-Trenaunay-Weber syndrome and Sneddon's syndrome. Acquired conditions include bartonellosis or HIV infection. In addition, surgery or injury could also cause hemangiomatosis. Currently, the understanding of hemangiomatosis is based on hemangiomatosis cases on the trunk and limbs because scalp hemangiomatosis rarely occur. A litera-

Figure 6. The scalp healing process. A. The scalp was sutured after excision of the mass. The scalp was aligned well and showed no defects. B. Scalp healing was poor 15 days after surgery, resulting in scalp defects. C. One month after surgery, a large portion of the scalp was growing well, and only the incision site exhibited poor healing. D. Two months after surgery, the scalp healed well.

ture review of hemangiomatosis retrieved only one case of s scalp hemangiomatosis reported by Kayaselcuk et al. in 2002. Infection by rod-shaped bacteria was found in lesion tissue of the hemangiomatosis in this patient. The cause was diagnosed as bacterial infection. Due to small size of the lesion and its localization inside the scalp, the lesion was completely excised with little difficulty [3].

Scalp hemangiomatosis can become very large, as reported in this case. The patient reported in this study was a young female. The scalp hemangiomatosis grew for 14 years without any specific cause. Previous pathological examinations did not show inflammation induced by the bacteria in the tissue. The lesion was small and soft and had not been treated. Over the 14 years, the hemangiomatosis had gradually grown along the scalp and finally involved a large portion of scalp and formed a large lesion, resulting in the ulceration at the center of the mass and scar formation. Although the hemangiomatosis case reported in the

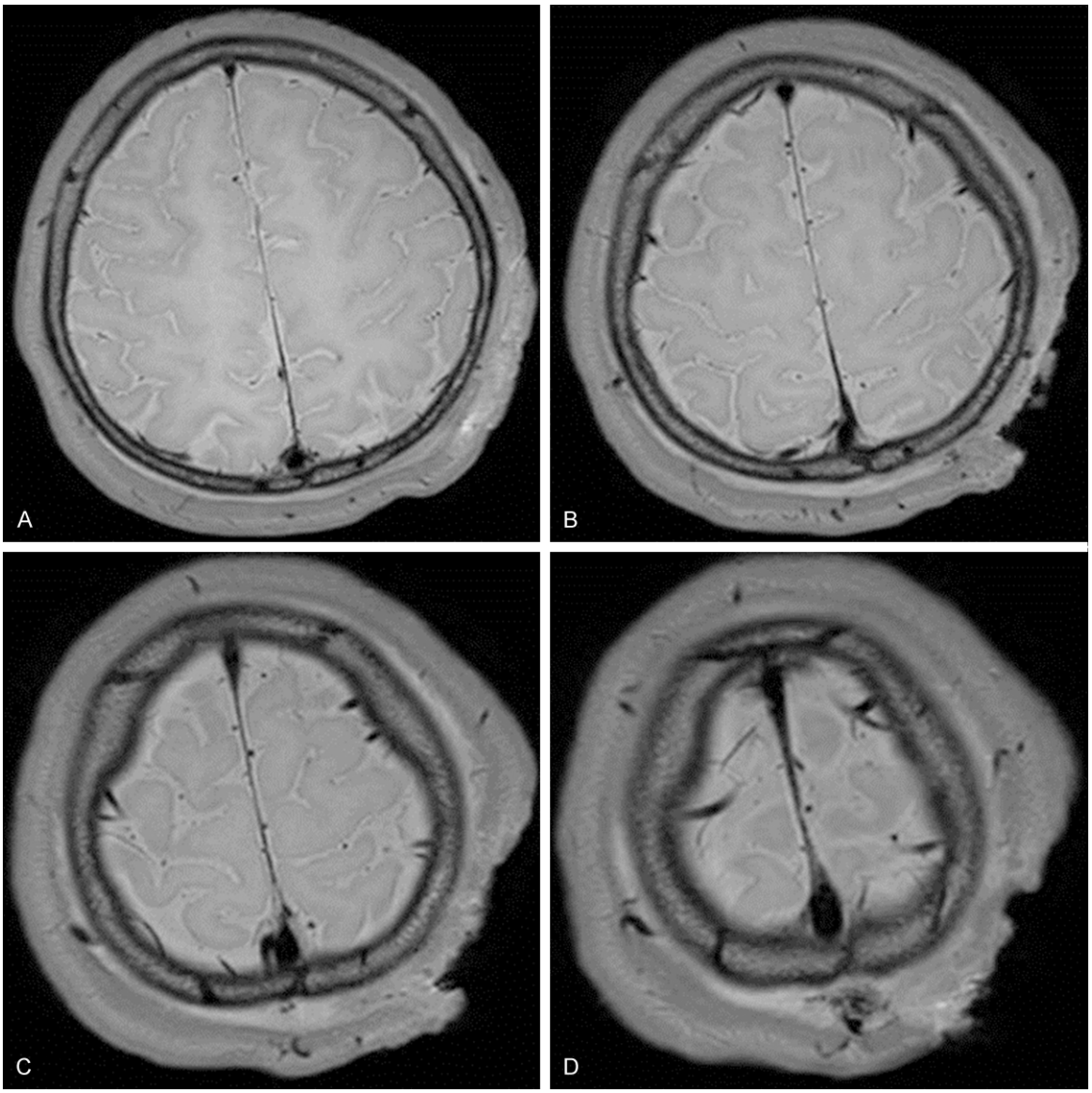

Figure 7. Re-examination of the head by MRI 20 days after surgery. A-D. Different MRI sections show that the lesion was completely excised. Scalp defects were observed locally, and the lumen of the expanding arteries in the tissues surrounding the lesion had recovered.

present paper exhibited growth over a large area of the scalp and bulging above the scalp, medical imaging examinations and surgery confirmed that the lesion had not invaded the skull or exhibited spreading and extensive growth. Because there are very few reports of scalp hemangiomatosis, it is not known whether scalp hemangiomatosis tend to invade the skull and exhibit vertical growth. Invasion of the skull and dura will make the treatment of hemangiomatosis extremely difficult.

The appearance and medical imaging characteristics of scalp hemangiomatosis are similar to those of scalp artillery and venous abnormalities; therefore, differentiation is required. Congenital scalp hemangiomatosis seldom occur alone; they are often accompanied by other syndromes such as Klippel-Trenaunay-Weber syndrome and Sneddon's syndrome. Acquired scalp hemangiomatosis are typically associated with surgery or injury [5]. Scalp artery and vein abnormalities do not have these characteristics and mainly manifest as vascular malformation clusters. The pervasive growth and appearance of the patched redness on the surface that occurs in hemangiomatosis is rare for these abnormalities [6]. However, the imag-

ing characteristics of the hemangiomatosis in this case were very similar to those of artery and vein abnormalities, e.g., abnormal malformation clusters, obvious blood supply arteries, and draining veins shown by MRI and DSA. However, the present study demonstrated rich adipose tissues and penetration of many abnormal blood vessels in the mass, which is different from the structure of artery and vein abnormalities.

Due to the above pathological changes of scalp hemangiomatosis, the blood vessels do not easily contract during surgery, resulting in bleeding. To reduce the bleeding, ultrasound and DSA examinations were used to locate the main blood supply arteries, which were then cut and ligated. At the same time, sutures were placed around the lesion to constrict the blood supply. All of these procedures ensure the successful excision of the lesion. The above method for the excision of hemangiomatosis can effectively reduce the blood supply. The poor healing and necrosis of the scalp after surgery could be related to the extension of the hemangiomatosis and its effect on the healing ability of the surrounding normal scalp tissues. An MRI examination was performed after surgery and clearly revealed the internal structure of the blood flow-enriched lesion and excluded the interference of other tissues [7]. The MRI findings showed that the excision was complete, and the diameter of the blood supplying arteries surrounding the lesion had returned to normal, which could be due to the decrease in blood pressure resulting from the decreased blood flow. Because hemangiomatosis tend to relapse, this case is still being followed up.

In summary, large hemangiomatosis can occur on the scalp and require sufficient preoperative evaluations, including the necessary examinations by ultrasound and DSA, prior to the treatment. After locating and ligating the feeding arteries, suturing the tissues around the lesion should also be performed to effectively control bleeding during surgery. If possible, the lesions should be completely excised to expose the healthy scalp. If scalp defects occur, frequent changes in medication and iodoform gauze packing of the defective area can effectively promote healing.

Disclosure of conflict of interest

None.

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