A Case of Venous Anomaly of Diploic Origin Successfully Treated by Preoperative Direct Puncture Sclerotherapy

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Objective: One case of venous anomaly arising from the cranium in which intraoperative hemorrhage could be effectively controlled by direct puncture sclerotherapy using N-butyl-2-cyanoacrylate (NBCA) is reported.

Case Presentation: A 76-year-old male noted gradually progressing right exophthalmos and swelling of the right temporal region and was found by imaging studies to have a space-occupying lesion extending inside and outside the cranium and in the orbit. On diagnostic biopsy of the extracranial part under local anesthesia, massive and spouting venous hemorrhage was observed. From intraoperative and pathological findings, a diagnosis of venous anomaly was made, and sclerotherapy was performed using NBCA under direct percutaneous puncture. After 4 days, the lesion could be resected totally with only slight bleeding.

Conclusion: Sclerotherapy using NBCA by direct puncture is considered useful as a preoperative treatment for venous anomalies.

Keywords ► venous anomaly, N-butyl-2-cyanoacrylate, direct puncture, sclerotherapy

Introduction

Venous anomaly has been described as a slow-flow blood-pooling lesion with venous lumens dilated in a cavernous or cystic pattern resulting from dilation of venous components due to hypoplasia of vascular endothelial cells in the course of embryonic vascular development. It may affect any part of the body including the face, trunk, and limbs, but is observed most frequently in the head and neck regions. It also arises in bones and abdominal organs as well as the skin and soft tissue.1) Small asymptomatic venous anomalies are often observed without treatment, but sclerotherapy is considered the first-line treatment for superficial large symptomatic lesions, particularly those in which blood is pooled in the lumen (lesions conventionally called cavernous hemangioma).2,3) However, for lesions in deep areas, such as intracranial and orbital lesions and those in which mass effect poses a problem, surgical resection is a therapeutic option, but total resection is often difficult due to a high risk of bleeding.4)

In this study, we obtained a satisfactory outcome in a patient with venous anomaly that arose from the diploe of the temporal region and extended in a complicated pattern to extracranial, intracranial, and intraocular regions by surgical resection after reducing the blood flow in the lesion by transarterial embolization and percutaneous direct puncture sclerotherapy. This case is reported.

Case Presentation

The patient was a 76-year-old male who complained of exophthalmos of the right eye and swelling of the
right temporal region. He had histories of nasal polyposis (surgically resected), mild bilateral deafness, and renal cysts. There was no particular familial history.

The patient had noted exophthalmos of the right eye and swelling of the right temporal region a few months before his consultation with a local doctor due to gradual exacerbation. Since abnormalities were detected by imaging examinations, he was referred to our department.

On neurological and physical examinations, no clear neurological deficits, including visual dysfunction such as diplopia were noted. In the swollen area of the temporal region, there was no color change in the skin, and no pulsation or vascular murmur was detected by palpation or auscultation. The swollen area was highly elastic and showed slight enlargement during the Valsalva maneuver.

On blood tests, no abnormality was noted in general chemistry or clotting items.

Concerning imaging examinations, head CT showed destruction of the temporal bone and upper wall of the orbit and extracranial, intracranial, and intraorbital spread of the lesion. Since the internal and external tables of calvaria were spread apart from the diploe, thinned, and ruptured, the lesion was considered to have arisen from the diploe (Fig. 1). No phlebolith was observed.

MRI of the head delineated the lesion as a hypointense mass on T1-weighted imaging, heterogeneous hyperintense mass on T2-weighted imaging, and multilocular mass on gadolinium contrast-enhanced imaging due to heterogeneous contrast enhancement (Fig. 2). On cerebral angiography, the lesion was markedly contrasted due to feeding from the anterior deep temporal and middle meningeal arteries, but no feeding artery from the internal carotid system was found (Fig. 3). Whole-body fluorodeoxyglucose (FDG)-positron emission tomography (PET) imaging showed no abnormal accumulation (image not shown). Ultrasonography was not performed.

Treatments

Biopsy: For histological examination, biopsy was performed in the extracranial part of the lesion by making a small incision in the scalp immediately above the lesion under local anesthesia. When the temporal muscle was incised, a dark red lesion covered by the periosteum was exposed. The structures to the periosteum were normal, and the periosteum, which was displaced by the lesion, was elevated and thickened. Incision of the surface of the lesion caused vigorous spouting venous bleeding. The interior of the lesion was hollow, and only a small amount of tissue could be collected with forceps, but swelling of the temporal region was slightly reduced after biopsy. Intraoperative and pathological findings suggested venous anomaly. Since the lesion spread into the orbit and caused marked exophthalmos, the condition was considered an indication for surgery.

Preoperative sclerotherapy: Since the lesion was found by biopsy to be at a high risk of bleeding, preoperative...
sclerotherapy was planned. A microcatheter was inserted serially into the anterior deep temporal and middle meningeal arteries, which were feeding vessels, and when they were embolized with 20% N-butyl-2-cyanoacrylate (NBCA), contrast-enhancement of the lesion disappeared on angiography (Fig. 3). When the lesion was palpated from over the scalp, the mass retained tension and protruded as before embolization, and the blood flow was not considered to have been sufficiently reduced to the interior of the lesion. The lesion was punctured percutaneously using an 18G indwelling needle, and the contrast agent was slowly injected. Since the contrast agent widely diffused in the lesion, embolization of the interior was judged to be insufficient. When, as additional sclerotherapy considered in advance, 12.5% NBCA was injected very slowly through an indwelling needle, which was inserted percutaneously, it spread slowly and diffusely like rime on trees in the three compartments (extracranial, intracranial, and intraorbital) (Fig. 4). A total of 13 mL was injected. During the injection of NBCA, continuous compression of the cervical veins and intermittent Valsalva’s maneuver were performed in combination. No efflux of NBCA through the draining vein was observed. No abnormality of the visual function was noted after sclerotherapy.

Surgery: Surgical resection of the lesion was performed 4 days after embolization. NBCA has diffused to all regions of not only the extracranial part, which was punctured, but also the intracranial and intraorbital parts, and the interior of the lesion was completely thrombosed. Unlike the time of biopsy, bleeding from the lesion was very slight (Fig. 5). The entire circumference of the lesion was clearly demarcated from the surrounding structures, the entire intracranial
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Fig. 3  Angiography. Frontal images of right internal carotid angiography (A) arterial phase, (B) capillary phase, arterial phase images of right external carotid angiography (C) frontal, (D) lateral, capillary phase images of right external carotid angiography (E) frontal, (F) lateral. Arterial phase images after embolization of the feeding arteries (G) frontal, (H) lateral. On right internal carotid angiography, an avascular area (B) broken line was observed in the area corresponding to the lesion. On right external carotid angiography, influx of blood from the middle meningeal and anterior deep temporal arteries was observed. The arrow and arrowhead (H) indicate the embolized middle meningeal and anterior deep temporal arteries, respectively.

Fig. 4  Fluoroscopic images during sclerotherapy by percutaneous puncture. The images in the upper (A) and lower (B) rows are arranged in order of time from left to right. In both rows, the images to the extreme right are those after sclerotherapy. NBCA injected percutaneously are seen to gradually diffuse to deeper compartments of the lesion. NBCA: N-butyl-2-cyanoacrylate
part of the lesion was located extradurally, and the intraorbital part was located outside the cone of the extraocular muscles, so the entire lesion could be resected with ease.

Pathological findings
Since a cluster of markedly tortuous and dilated blood vessels was observed, and no abnormal proliferation of endothelial cells was noted, a diagnosis of venous anomaly was made. NBCA permeated all parts of the lesion, and strong thrombus formation was confirmed in its interior (Fig. 5).

Postoperative course
Total resection of the lesion could be confirmed by postoperative MRI (Fig. 6), and exophthalmos was resolved. No visual dysfunction, such as reduced visual acuity, narrowing of the visual field, or diplopia, was noted after surgery.

Discussion
Lesions called hemangiomas including vascular tumors and vascular anomalies were classified into hemangiomas and vascular anomalies in 1982 by Mulliken and Glowacki.5) Thereafter, the International Society for the Study of Vascular Anomalies (ISSVA) classified lesions into vascular tumors and vascular anomalies and established the current standard international classification.6) Cavernous hemangioma, venous hemangioma, intramuscular hemangioma, and synovial hemangioma, which used to be regarded as hemangiomas, are classified as venous anomalies by the ISSVA classification. Although there is still some confusion in terminology, it is worth reviewing the past literature on cavernous hemangioma by redefining it as venous anomaly.

There have been a number of reports that cavernous hemangioma arising from the cranium is a very rare disorder among bone neoplasms,1) but there are also reports that cavernous hemangioma is the most frequent lesion next only to osteoma if the search is limited to primary masses of the cranium.7,8) Thus, primary tumor masses of the cranium are rare, but "cavernous hemangiomas," or venous anomalies, account for a large percentage of primary tumor masses of the cranium, and it is necessary to include them in differential diagnoses.

Sclerotherapy is the first-line treatment for superficial venous anomalies.2,3) However, for venous anomalies in deep areas, such as the cranial base and orbit, en bloc resection has been regarded as the standard treatment since the report by Cushing in 1923.9) In addition, as direct puncture...
of the deep lesion for sclerotherapy is difficult, there have been a number of reports of surgical resection. Particularly, in the orbit, the mass effect is the greatest problem, and priority has been placed on surgical resection. Venous anomalies are easy to bleed, and massive intraoperative bleeding interferes with total resection depending on the site and size of the lesion. A combination of surgical resection with sclerotherapy, which is safe and reliable, is necessary in some patients.

Generally, contrast enhancement of the lesion or delineation of the feeding artery is not considered to be observed on angiography of venous anomalies. However, concerning venous anomalies arising from the cranium (originally termed as “cavernous hemangiomas”), there have been reports that they were fed by the middle meningeal and superficial temporal arteries\(^{10,11}\) and that the lesion was contrasted on angiography in about half the patients.\(^{12}\) Venous anomalies arising from the cranium may have different properties compared with those arising from other sites. In the present case, also, the venous anomaly is considered to have originated from the diploe and was fed by the middle meningeal and anterior deep temporal arteries.

The interior of venous anomaly comprises wide vascular lumens partitioned by thin walls and sinusoidal lumens divided by epithelial monolayers and is filled with blood.\(^{13,14}\) Therefore, the percentage occupied by vascular lumens in the lesion is overwhelmingly large compared with tumors such as meningioma. This sinusoid structure is clearly visualized on angiography by direct puncture as emphasized by Kirchhoff et al.\(^{12}\) Therefore, sclerotherapy by direct puncture is the optimal method to deliver the sclerosing agent to all parts of the lesion and, indeed, is the first choice for the treatment of venous anomalies. In the present case, feeding arteries sufficiently large for catheter insertion were present, but, even if the embolic agent has been injected transarterially via these routes, the feeding arteries are occluded before it is sufficiently diffused in the large sinusoid space. Although there have been a few reports of the usefulness of transarterial embolization for venous anomalies,\(^{15}\) its effectiveness remains questionable. Particularly, for lesions with complex multilocular morphology as the one in the present case, it is important to ensure permeation of the sclerosing agent injected by direct puncture to all parts of the lesion. In sclerotherapy for venous anomalies, sclerosing agents, such as ethanol, Oldamin, and polidocanol, are frequently used, but we selected low-concentration NBCA diluted with lipiodol in our patient. The selection was based on its good visibility, excellent diffusibility in the lesion, our familiarity with its properties, and the absence of the problem of retention of the sclerosing agent in sclerotherapy performed before surgical resection.

Technically, it is necessary to devise measures to reduce the blood flow in the lesion and slowly inject NBCA while preventing its entry into the draining vein. The use of low-concentration NBCA to delay its hardening and facilitation of diffusion of NBCA by warming lipiodol used to dilute it to reduce the viscosity are considered to have been useful. Since the blood flow velocity is lower in venous anomalies than in arteriovenous malformations, the risk of unintended entry of the embolic agent into the draining vein is considered to be relatively low. In the
present case, however, we first occluded the feeding arteries and applied continuous compression of cervical veins and intermittent Valsalva maneuver by the cooperation of the patient. We have the impression that these techniques were useful for the prevention of entry of the embolic agent into the draining vein, but its validation is difficult by the experience of this case alone. Low-concentration NBCA diluted with lipiodol had excellent visibility, and its injection rate could be adjusted by real-time monitoring of its diffusion. NBCA has very high thrombogenicity even at a low concentration of 12.5%, and it showed sufficient effects such as the absence of bleeding from the lesion in surgical resection. These results suggest that the advantages of the selection of NBCA as the sclerosing agent could be fully utilized.

The present case exhibited marked exophthalmos due to the intraorbital lesion and required surgical resection. However, the lesion had complex multilocular morphology and occupied a wide space in the cranium, and, in consideration of the state of bleeding at biopsy, the possibility of massive bleeding would have been high without preoperative sclerotherapy, probably making resection of the intraorbital lesion impossible. Since part of the lesion had destroyed the external table in this case, the lesion could be approached percutaneously, and preoperative sclerotherapy was an effective choice.

## Conclusion

We encountered a case of venous anomaly arising from the diploe in which preoperative sclerotherapy was very effective. Since the lesion extended extracranially through a bone defect, sclerotherapy by direct percutaneous puncture was possible before surgery, and, although the lesion was a large multilocular venous anomaly involving the orbit, it could be completely resected safely with little bleeding. Sclerotherapy using low-concentration NBCA may be useful as a procedure before surgical resection.

## Disclosure Statement

Neither the first author nor any of the coauthors have any conflicts of interest to disclose regarding this paper.

## References