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Treatment of Galen Vein Arteriovenous Malformation with Gamma Knife Surgery

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Abstract:

The vein of Galen arteriovenous malformation is a rare abnormality of the fetal cerebral vascular system. It is caused by the persistence of a cerebral arteriovenous fistula of the median prosencephalic vein. Due to the high mortality rate of the surgical option in its treatment, the approach with embolization and radiosurgery combinations continues to be popular. We present a 3-year-old child patient who was accidentally detected by vein of Galen malformations during a screening for headache complaints with known heart disease.In the treatment of this patient, we performed radiosurgery therapy alone, rather than a combination. Our case is the third case of a and the youngest example child who was treated with only radiosurgery without embolization. Gamma knife therapy, which is a more convenient and effective treatment plan, is a method that can be safely used instead of open surgery with a high mortality rate.

Key Words:

Arteriovenous malformation, Galen vein, gamma knife, radiosurgery

Key Message:

Arteriovenous Malformations of the Galen Vein are quite difficult pathologies to treat, and many treatment modalities are possible. The multidisciplinary treatment approach is the most preferred. This patient, who was treated with a less invasive method such as Gamma Knife radiosurgery, may have paved the way for a treatment that can be tried at the first stage in terms of mortality and morbidity risk.

Vein of Galen malformations (VGOMs) are a rare cerebral intracranial vascular malformation.[1,2] VOGMs account for <1% of all vascular malformations, while they account for about 30% of pediatric cerebrovascular malformations.[1] The origin of VGOM is not entirely clear. It is characterized by primitive choroidal arteries and a primitive precursor of midline cerebral venous structures median prosencephalic vein of Markowski a precursor to the vein of Galen, between 6 weeks and 11 weeks of gestation.[3] It is usually manifested by secondary multiorgan failure due to excessive circulatory loading and the formation of the intracranial shunt during the neonatal period.

We present a case of a Galen vein arteriovenous malformation (AVM), which was detected asymptomatically despite being clinically worse in the childhood age group and treated using only radiosurgery modality. Our case is the third case of a child who was treated with only radiosurgery without embolization as a result of a literature review and received good outcomes.

Materials And Methods

Literature review

Literature search (PubMed database, 1892–2022) completed to include the following keywords: "Gamma Knife Surgery," "Cerebral arteriovenous malformations," "Vein of Galen malformation," and "embolization." Duplicate works were excluded.

Case: A patient child of 3 years old was diagnosed with VGOM in MRI images taken during etiology research due to headache complaints [Figure 1]. The dimension of the AVM was 16 × 16 × 12 mm with the thalamoperforators originating from the right vertebrobasilar system as seen in digital subtraction angiography (DSA). A thick varicose vein was draining to confluens sinuum and

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How to cite this article: Kuzucu P, Sözer A, Börcek AÖ. Treatment of Galen Vein Arteriovenous Malformation with Gamma Knife Surgery. Neurol India 2024;72:1092-4.

Submitted: 06-Oct-2022 **Revised:** 03-Jun-2023
Accepted: 04-Jun-2023 **Published:** 19-Oct-2024

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transverse sinus. Initial imagings show no evidence of previous bleeding. It was not suitable for embolization in its natural state due to the fact that our patient was found to be incidentally and that VGOM had thalamoperforators.

Results

18 Gy prescription dose and 50% isodose were used in Gamma Knife Surgery (GKS). AVM volume is measured as 4.4 cm^3 [Figure 2] five months later. The minimum dose AVM received was 9.9 Gy, and the maximum dose was 37.2 Gy [Figure 2]. Treatment was 98 min in total and performed under general anesthesia. The Gamma Knife model perfexion was used in the case. After the procedure, the patient is discharged without any additional sequela on the same day. The patient was under follow-up for 3 years, as a result of the DSA, VGOM was significantly reduced with approximately 1 cm of the residual nidus, which was mainly drained of blood from the posterior choroidal artery and some branches of the superior cerebellar artery [Figure 3]. Follow-up did not develop any new deficiencies and no bleeding was observed.

Discussion

The vein of Galen aneurysm malformation is a rare cerebral intracranial vascular malformation characterized by a cerebral arteriovenous shunt provided by various choroidal arterial supply vessels. For the first time, VGOM was described by Jaeger *et al*. in the middle of the twenty century.[4] Clinically, they usually occur with hydrocephalus and shunt fractions secondary to high-output heart failure or venous obstruction.[5]

At first, surgical approaches to fistula connections were tried in the treatment of VOGM, but since their results in very high rates of perioperative mortality so, the search for new treatments began. Therefore, the method of treatment with endovascular embolization, which began in the late 1980s, has become the standard of therapeutic care, which reduces mortality to about 15% and greatly reduces neurological morbidity. But now, treatment approaches are preferred, especially in combination with radiosurgery and endovascular therapy.[6] Berenstein *et al*.[7] as a result of their study reduced the neonatal mortality rate from 50% to 11% in less than 2 years by endovascular treatment.

Triffo *et al*.[8] reported they described radiosurgery practices in two of their cases. The first patient after unsuccessful embolization received gamma knife treatment and left a very small amount of residue during follow-up. The second patient performed only radiosurgery treatment after the malformation was not suitable for embolization in its natural state, and as a result, they observed complete closure in an adult patient. In every two cases, patients were followed without additional neurological deficits.[8]

According to a study of the results of radiosurgery treatment on VGOMs conducted by Payne *et al*.,[9] they reported a total of six cases. Three of them were mural type, while three were choroidal type VGOM. According to this study, a patient that is treated with a combination of GKS and embolization, and a patient that is cured after three failed embolization with GKS, two patients with primary GKS treatment at the age of four and six, and finally one patient who underwent GKS as a result of failed embolization four times were presented [Table 1]. Except for the last patient, they received positive responses from the treatment.[9] Our case is the third case of a child who was treated with pure radiosurgery without embolization. We represent the youngest example of successful radiosurgery treatment in childhood.

Conclusions

Due to the high probability of progressive neurological disability and the high mortality rate in open surgery, it makes it strong to include radiosurgery as a reliable option in the treatment algorithm for these complex lesions.

Table 1: Child patients treated with gamma knife in literature

Studies	Age	Type
Payne *et al.*	4	Choroidal
	6	Choroidal
Our case	3 years 2 month	Choroidal

Our case is the third case of a and the youngest example child who was treated with only radiosurgery without embolization

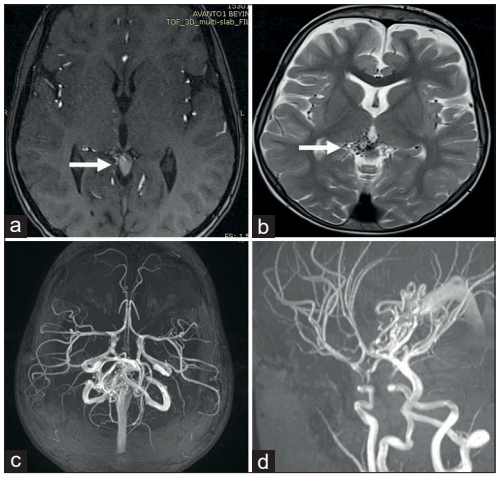


Figure 1: (a) Initial contrast-enhanced MR image showing a large aneurysmal Galen sac (arrowhead). (b) VOGM image in axial T2 MR sequence (arrowhead). (c) Preoperative VOGM image in axial angiography MRI sequence. (d) Preoperative VOGM image in sagittal angiography MRI sequence

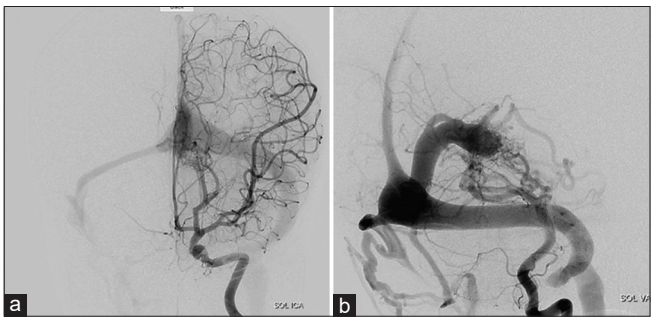


Figure 2: (a) VGOM image in marked DSA made before gamma knife surgery treatment planning. (b) An MRI and DSA image of treatment plans in GKS in the perfection model



Figure 3: (a) Follow-up image with selective angiogram of the postoperative oblique left internal carotid artery. (b) Follow-up image after 3 years, lateral selective angiogram of the left internal carotid artery. (c) Image of the left vertebral artery DSA

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Acknowledgment

We certify that the content of this manuscript, in part or in full, has not been submitted to any other journal in any form, and its publication has been approved by all co-authors.

Patient's consent

A full and detailed consent from the guardian has been taken. The patient's identity has been adequately anonymized. If anything related to the patient's identity is shown, adequate consent has been taken from the guardian. The journal will not be responsible for any medico-legal issues arising out of issues related to patient's identity or any other issues arising from the public display of the video.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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