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Case Report

Vein of Galen malformation with Obstructive Hydrocephalus- Endovascular Approach of Management : A Case Report

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Abstract:

The vein of Galen malformation is a rare arteriovenous shunt that is estimated to represent 30% of all pediatric vascular malformations. Less than 1% of cerebral vascular malformations are aneurysmal abnormalities of the vein of Galen, making it a rare disorder. Despite being uncommon, VGM is of particular clinical importance since the large cerebral shunt volume causes a significant mortality rate when left untreated because to date, the treatment of patients with VGM remains challenging. Here we report a case of an 11-month-old baby who had VGM with obstructive hydrocephalus and was effectively treated by transarterial glue embolization of the fistula and the venous sac. Subsequently, the baby made a complete recovery.

Keywords: vascular malformation; Vein of Galen malformation; endovascular treatment; embolization; Intracranial Vascular Malformation, Arteriovenous Malformation, transarterial glue embolization.

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Introduction:

Vein of Galen Malformation (VGM) is a congenital intracranial vascular anomaly1.The majority of Vein of Galen Aneurysmal malformations (VGAMs) become symptomatic in the neonatal period and if left untreated have an almost 100% morbidity and mortality2. They can be divided into two types: mural and choroidal and they can manifest as cardiac congestive heart failure at the neonatal period, renal failure, cerebral ischemia and hydrocephaly3. Hydrocephalus is the most common finding associated with a diagnosis of VOGM4. The choroidal type is the most severe form of VGM, which frequently presents with high-output cardiac failure, macrocephaly with loud bruits, and dilated orbital veins due to multiple high-flow fistulas with less restriction of outflow5. Treatment

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options for VGMs have historically included either open surgery, a direct transtorcular approach to ligation, transarterial embolization, or transvenous embolization. Reports show nearly 100% mortality with open surgical approaches; transtorcular approaches have, likewise, not been successful; and transvenous approaches are reported to have poor cognitive outcomes6. The first attempt to treat a VGM was at the beginning of the last century, wherein bilateral internal carotid artery ligations were performed on an infant who had presented with intracranial hypertension7. At present, endovascular embolization is preferred for treating VGMs89. The embolization is performed by transarterial, transvenous or combined approach, using coils, cyanoacrylate or Onyx glue10. The goal is to decrease the blood flow through the fistula reducing the venous hypertension and blood steal, which allows clinical improvement despite lack of total obliteration of the fistulas10. Complications of the procedure include cerebral hemorrhage, vessel perforation, hydrocephalus and sinus thrombosis, leg ischemia11.

In our case, an 11 months old male baby presented with seizure, decreased level of consciousness and hypertension hydrocephalic syndrome. The baby underwent transarterial embolization of the mural type malformation.

Case Report:

A 1 year old baby presented with repeated episodes of seizure since birth and delayed psychomotor development, and focal neurological symptoms and dilated superficial vein. MRI and MRV with contrast revealed Vein of Galen Malformation and Obstructive hydrocephalus with dysmaturation of sigmoid sinus and jugular vein with retrograde venous drainage. The baby underwent endovascular embolisation. Postoperative CT showed nearly complete embolization of VOGM. In the immediate postoperative period the baby made significant improvement. 2 months follow up CT revealed reduction of thrombosed VOGM and resolving Hydrocephalus. The baby made significant improvement. His GCS 15/15, playful, gaining all milestones of development normally with time.

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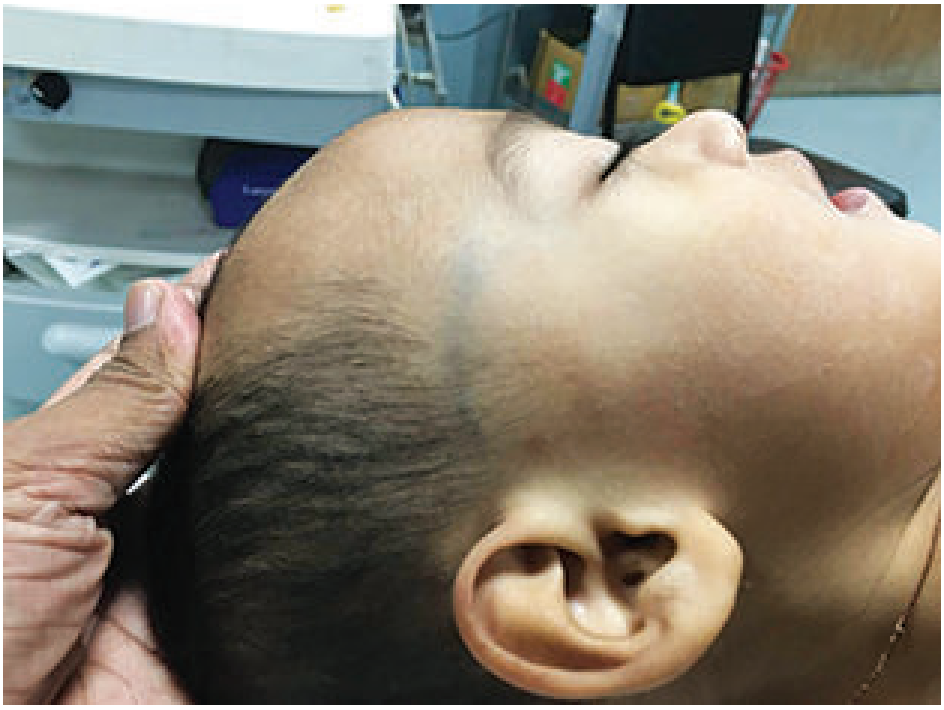

Fig.-1: *Dilated superficial vein*

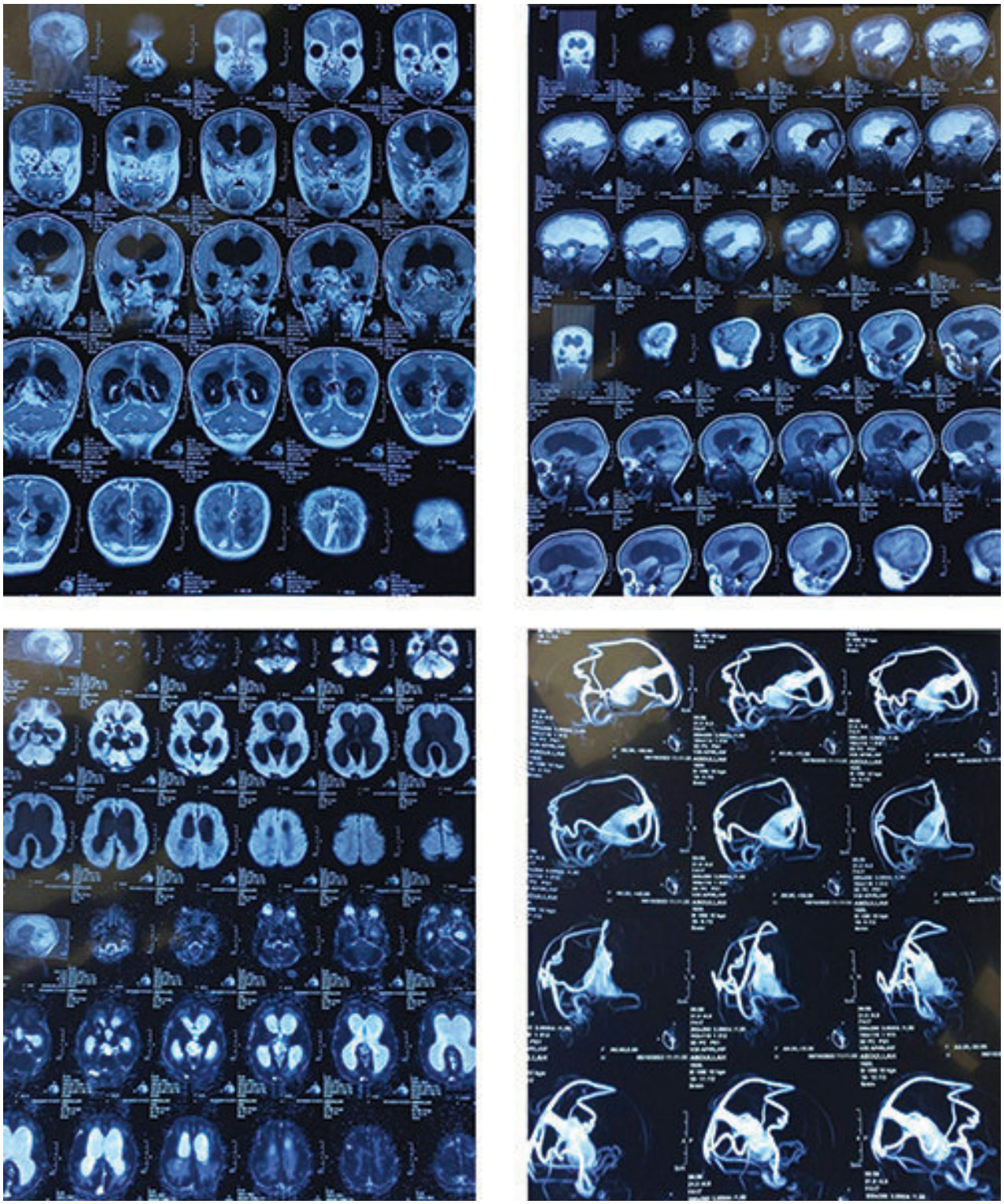

Fig.-2: (a,b,c, d): MRI and MRV with contrast reavealed Vein of dysmaturation of sigmoid sinus and jugular vein with retrograde venous drainage to Vein of Galen and Obstructive hydrocephalus with Vein of Galen Malformation.

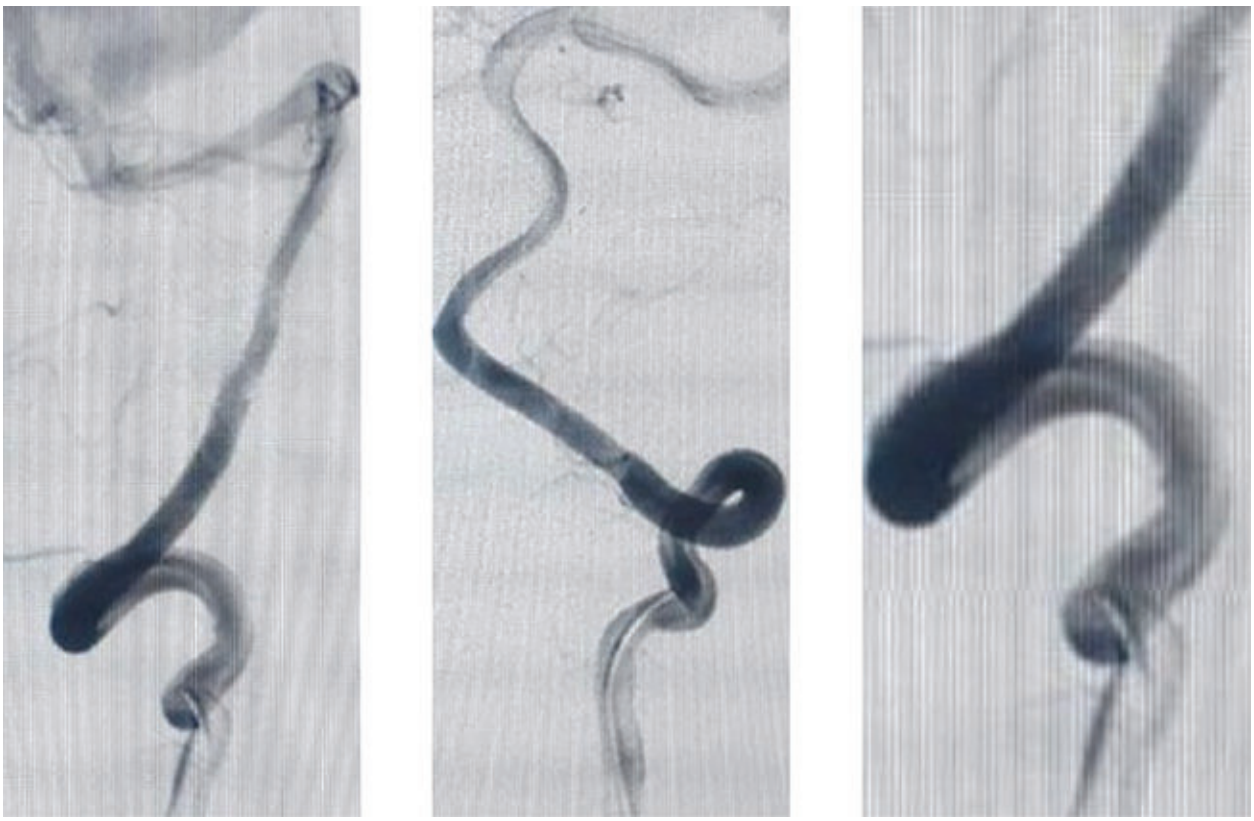

Fig.-3 (a,b,c): *Digital Subtraction angiography revealed mural type VOGM feeded by left posterolateral choroidal artery.*

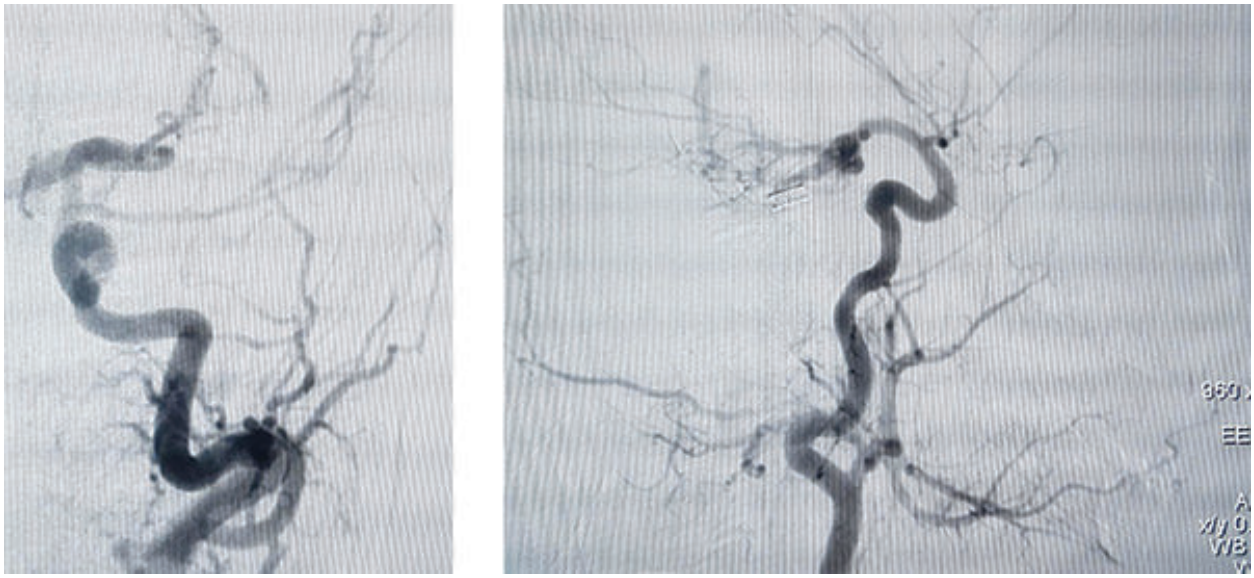

Fig. 4 (a,b): *Endovascular ernbollsatlon done by· n-butyl cryoacrylate of left posterolateral choroidal artery and VOGM.*

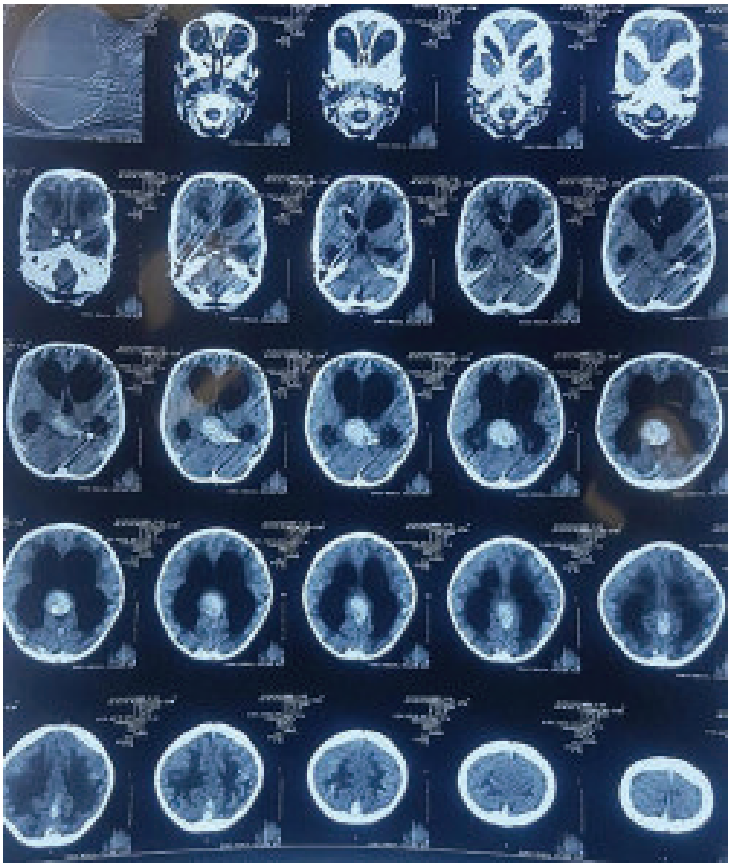

Fig.-5: *Immediate Postoperative CT shows thrombosed VOGM*

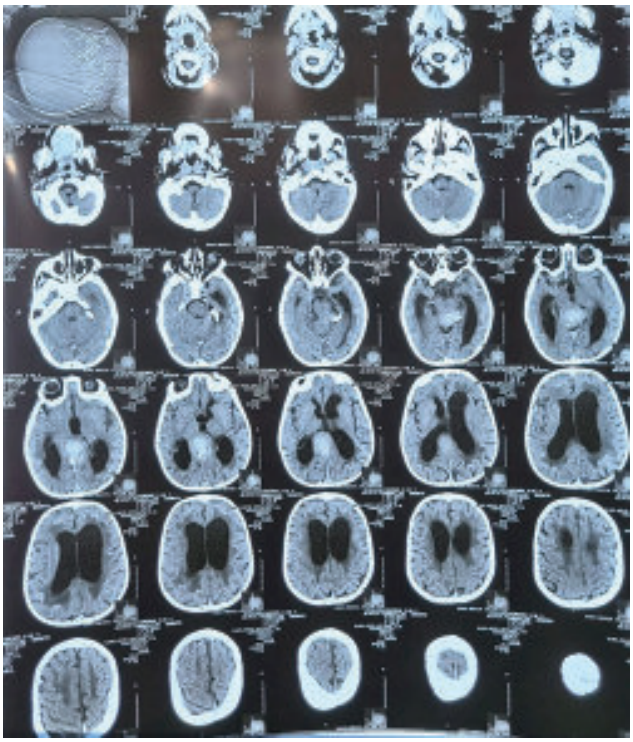

Fig.-6: *2 months follow up CT shows deceased size of thrormbosed VOGM and resolving Hydrocephalus*

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Fig.-7: *In 2 months Follow up visit- the baby appears to playful and started to achieve all milestones of devellopment.*

Discussion:

If VGM is diagnosed in childhood or adolescence and treated appropriately, life expectancy increases significantly [12]. Emphasis is then placed on treatment of hydrocephalus and recurrent seizures and prophylaxis of intracranial hemorrhage, with the goal of allowing normal or quasi-normal neurological development [13]. Persistent venous hypertension with malabsorption of cerebral spinal fluid and initiation of intracranial hemorrhage due to secondary changes in venous outflow (including persistence of fetal falciform sinus, lack of development of the rectus sinus, or dilatation of the internal cerebral veins) are pathophysiological explanations for the exacerbation of symptoms [14]. In our case, a 1 year old baby presented with repeated episodes of seizure since birth and delayed psychomotor development, and focal neurological symptoms and dilated superficial vein. MRI and MRV with contrast revealed Vein of Galen Malformation and Obstructive hydrocephalus with dysmaturation of sigmoid sinus and jugular vein with retrograde venous drainage.Regarding treatment, Neurosurgery as a first-line causal treatment is a thing of the past. Yasargil and colleagues have extensively engaged in microsurgical approaches, with notable success in the field. However, in a 2013 review article, Yasargil himself pointed out that neurosurgery in VGM Yasargil types IVa–c should be considered risky because of the involvement of the diencephalic and mesencephalic arterial feeders[15] In 2010,Jea et al. called for the use of endovascular embolization of the VGM to rebalance hydrovenous dynamics as the first-line option rather than directly managing the hydrocephalus by CSF diversion[16].The prognosis for children harboring these lesions has significantly improved over the last 3 decades: due to refinements in interventional endovascular techniques, it is now possible to obliterate the malformation with excellent results. Figures from the comprehensive literature review of Khullar et al.[17] indicate that as many as 84

% of children treated by endovascular techniques after the turn of this century may experience a good or fair outcome. It must be recognized, however, that the paper by Meila et al.[18] has the merit of adding exhaustive evidence to the neurosurgical literature showing that changes of ventricular size from enlarged to normal do occur after successful endovascular treatment. Here, we author opted Transarterial embolization of VGM and feeding vessel left posterolateral choroidal artery. The baby tolerated the procedure well. Postoperative CT showed nearly complete embolization of VOGM. In the immediate postoperative period the baby made significant improvements. 2 months follow up CTrevealed reduction of thrombosed VOGM and resolving Hydrocephalus. The baby made significant improvement. His GCS 15/15, playful, gaining all milestones of development normally with time.

Conclusion:

It is quite uncommon for children to present with both acute obstructive hydrocephalus and a deeply positioned, unruptured AVM. Although focal neurologic impairment, seizures, and headache are the three symptoms that are most frequently present, in certain situations, abrupt neurologic deterioration brought on by hydrocephalus may be the presenting symptom. Timely endovascular embolization can be a very effective modality of management in treating a baby with a similar presentation, like in our case.

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