Arteriovenous Malformation of the Vein of Galen Coexistent with a Straight Sinus Anomaly: The Role of Transcranial Doppler Ultrasound in Diagnosis and Postoperative Follow-up

Sinus Rektus Anomalisiyle Birlikte Görülen Galen Veni Malformasyonu: Tanıda ve Ameliyat Sonrası İzlemde Trankskraniyal Doppler Ultrasonografinin Yeri

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Abstract: The authors report a case of aneurysmal malformation of the vein of Galen associated with straight sinus anomaly (accessory straight sinus) in a 6-month-old infant presenting with clinical signs of hydrocephalus. The malformation was confirmed by computerized tomography, magnetic resonance imaging, cerebral angiography, and transcranial Doppler. The patient was operated on through a bilateral parieto-occipital craniotomy and interhemisferic approach on both sides of the falx without any complication. Postoperative angiography and transcranial Doppler investigation confirmed total occlusion of the fistulas.

Key Words: Hydrocephalus, infant, transcranial Doppler ultrasound, vascular malformation, vein of Galen

Özet: Hidrosefali bulguları olan 6 aylık bir bebekte sinüs rektus anomalisi *(sinus rectus accessorius)* ile birlikte Galen veninin anevrizmal malformasyonu sunulmuştur. Malformasyon bilgisayarlı tomografi, manyetik rezonans görüntüleme, serebral anjiografi ve transkraniyal Doppler incelemeleriyle gösterilmiştir. Hasta bilateral parieto-oksipital kraniotomi ve falksın her iki tarafından interhemisferik yaklaşım ile ameliyat edilmiş, ameliyat sonrası komplikasyonsuz seyretmiştir. Cerrahi sonrası anjiografi ve transkraniyal Doppler incelemeleri fistülün tamamen tıkandığını doğrulamıştır.

Anahtar Sözcükler: Çocuk, damarsal malformasyon, Galen veni, hidrosefali, transkraniyal Doppler ultrasonografi

INTRODUCTION

Aneurysmal malformation of the vein of Galen is a rare anomaly. This lesion is frequently supplied by an anomalous branch of the carotid and/or basilar circulation on both sides (2,4,5,7,20). In some cases, the straight sinus is absent, and venous flow is drained through an accessory straight sinus into an accessory confluens (20).

The diagnosis of this lesion is made by computerized tomography (CT), magnetic resonance imaging (MRI), magnetic resonance angiography (MRA), transcranial Doppler ultrasonography (TCD), and cerebral angiography (7,9,17). Although the treatment of this lesion is controversial modes of treatment are surgery, embolization or a combination of these two methods (3,5-7,10,19,20).

In this report, we present surgically treated type 2 galenic arteriovenous malformation (AVM) (20) in which we used TCD as a diagnostic tool. The

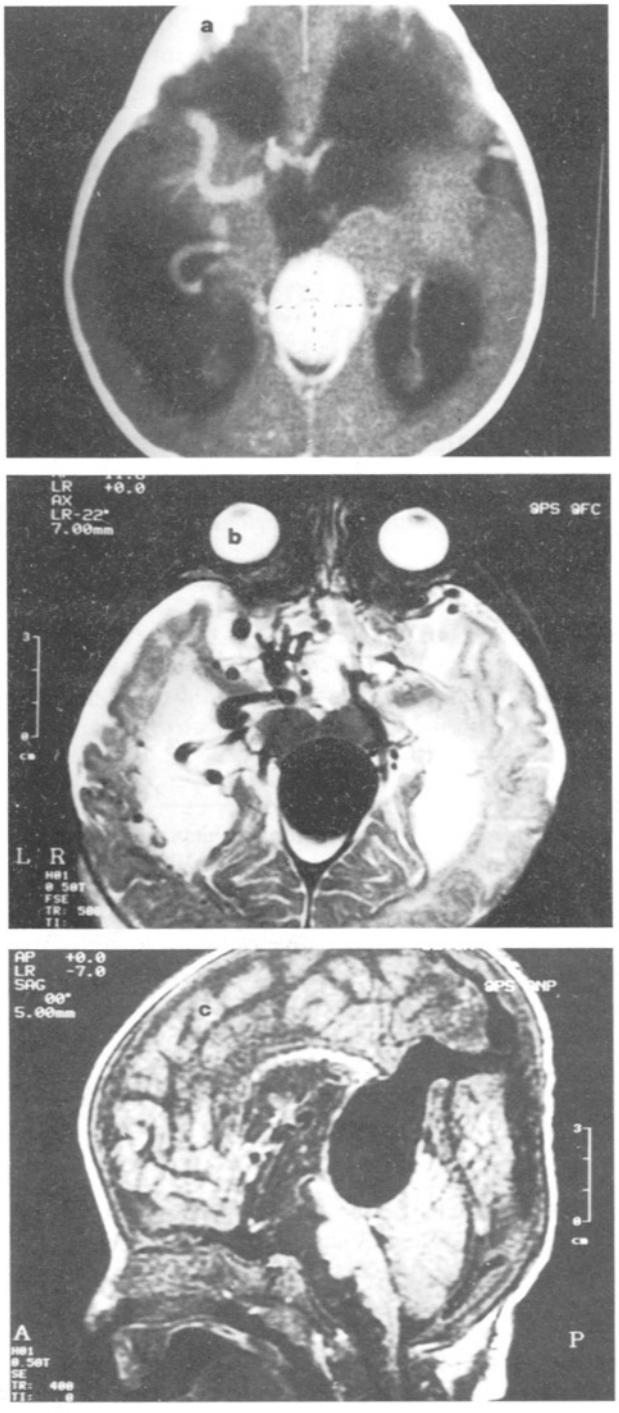

Figure 1, a) CT scan reveals a 30x32x40-mm enhancing mass in the pineal region, b) axial MR image demonstrates that the both PCAs feed a large aneurysmal dilation, c) sagittal MR image showing accessory straight sinus and stenosis of straight sinus.

importance of TCD investigation was discussed to determine the dominant feeders preoperatively and to confirm vascular occlusion in the postoperative period.

CASE REPORT

A rapidly increasing head circumference was noted in a 6-month-old infant 3 months after his birth. At 6 months, the head circumference was 51 cm. He was admitted to our hospital with the findings of hydrocephalus. Cardiac examination revealed no abnormalities. The chest x-ray was normal. CT scan revealed a 30x32x40-mm enhancing mass in the pineal region, indenting the posterior aspect of the third ventricle and symmetric obstructive hydrocephalus with dilated third and lateral ventricles (Figure 1). A ventriculoperitoneal (VP) shunt was inserted on June 23,1995. After the shunt operation, MR examination suggested a possibility of galenic AVM with an accessory sinus and stenosis of straight sinus (Figure 1). TCD investigation revealed high flow velocity (Vm 132 cm/sec), low pulsatility indices (PI 0.46) and decreased CO2 reactivity in the left posterior cerebral artery (PCA) (Figure 2), and slightly high flow velocity (Vm 51 cm/sec), slightly low pulsatility indices (PI 0.58) and decreased CO2 reactivity in the right PCA. TCD investigations were performed using a pulsed, range-gated 2-MHz TCD (model multi-DOP-X, DWL

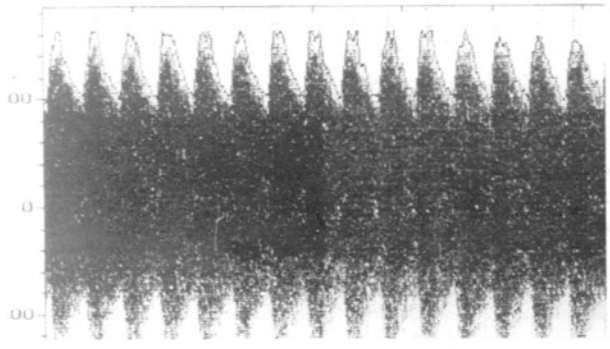

Figure 2. TCD investigation shows increased flow velocity (Vm) and decreased pulsatility indices (PI) values on the left PCA (normal values, Vm 37±7.6 cm/sec, PI 0.83±0.24).

Company, Überlingen, Germany). Four-vessel cerebral angiography demonstrated an aneurysmal dilation fed by numerous branches of both PCAs and posterior thalamic perforators (Figure 3). There was stenosis of the straight sinus and drainage was to the superior sagital sinus and to the accessory confluens through an accessory straight sinus.

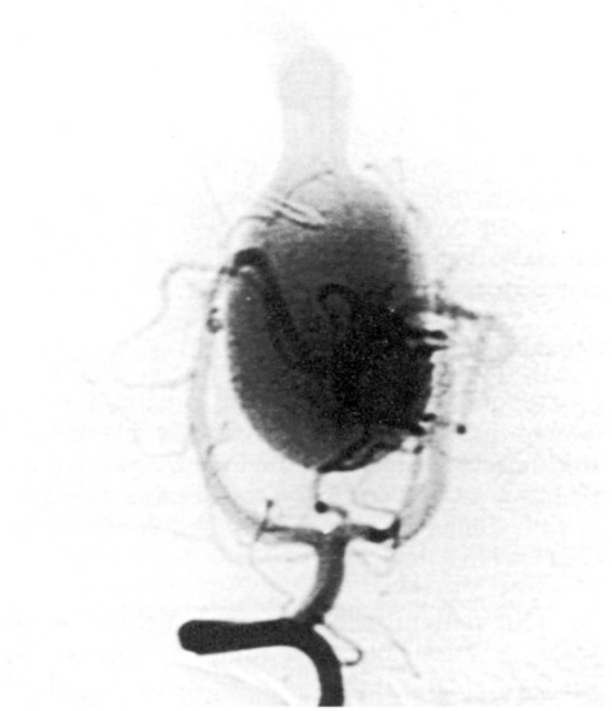

Figure 3. Cerebral angiography shows fistulous connections of both PCAs to the vein of Galen.

The patient was operated on July 3,1995 through a bilateral parieto-occipital craniotomy and interhemisferic approach on both sides of the falx in the sitting position. Feeders arising from both PCAs were coagulated and sectioned, but the sac was feeding dominantly from the left side. The wall of the sac was very thick. Careful application of the bipolar coagulation diminished the size of the aneurysm, permitting possible access to the small posterior thalamic perforators; these were coagulated and divided without difficulty. After the occlusion, the aneurysmal sac collapsed. The operation time was 5 hours; blood transfusion was 90 ml. There was no cardiovascular abnormality during operation.

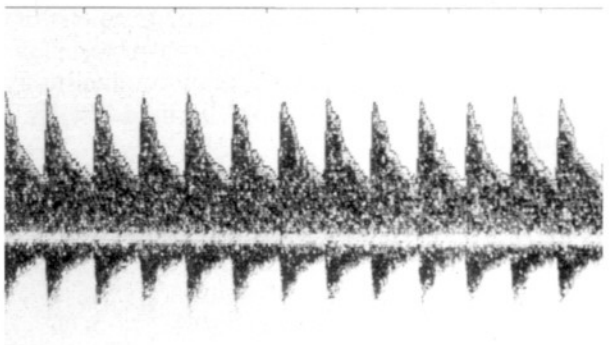

Figure 4. Postoperative TCD investigation shows normal values of Vm and PI on the left PCA.

The postoperative period was uneventful. A control TCD investigation revealed normal values of mean flow velocity (Vm 53 cm/sec), and pulsatility indices (PI 0.90) on the left PCA (Figure 4) and mean flow velocity of 48 cm/sec, pulsatility indices of 0.88 on the right PCA. Postoperative angiogram showed total obliteration of the malformation (Figure 5). The CT scan showed subdural fluid accumulation (Figure 6).

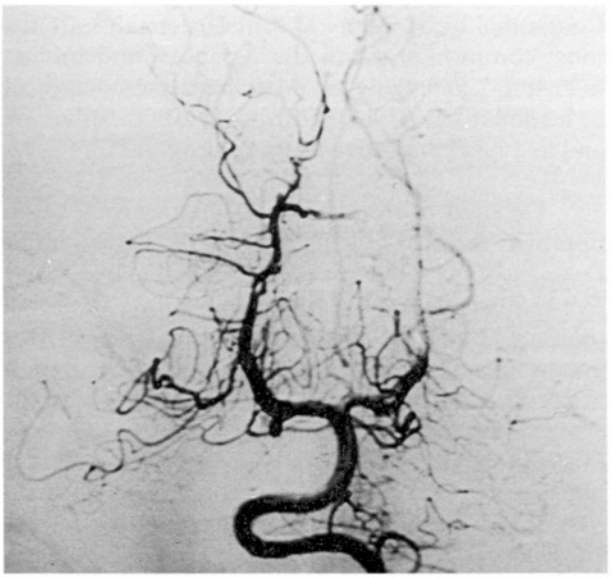

Figure 5. Postoperative angiography, 2 weeks later, shows the complete elimination of the vein of Galen.

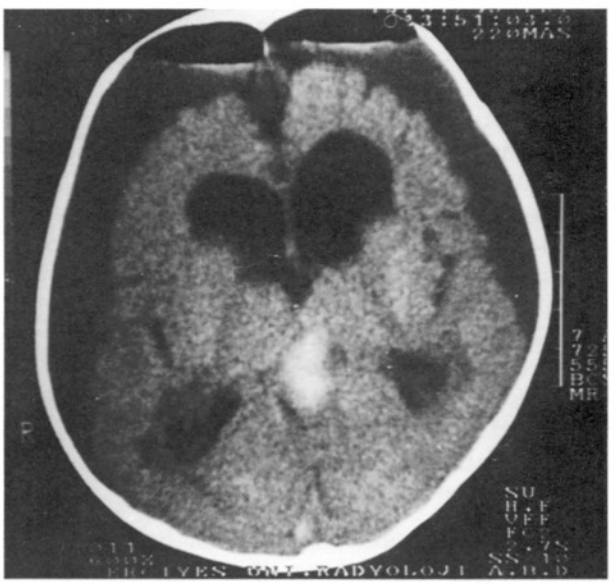

Figure 6. Postoperative CT scan shows the collapsed aneurysmal sac and subdural fluid accumulation.

DISCUSSION

The malformation or aneurysm of the vein of Galen may be defined as an aneurysmal dilation of the vein of Galen that has an arterial input from an anomalous branch of the carotid and/or basilar circulation on both sides (2,4,5,20). The anatomical and clinical aspects of the galenic AVM are now well established and classified by Yaşargil into four categories (20).

Clinical manifestations of the galenic AVM are related to the cardiovascular and nervous systems. Congestive heart failure and hydrocephalus are the most common signs in the newborn and infants (6,11,13,14). Especially in the newborn diagnoses must be made as early as possible in order to initiate therapy and to avoid congestive heart failure (1,4,7).

Detailed anatomical and dynamic analysis is obtained from CT, MR, MR angiography, color Doppler ultrasonography and cerebral angiography (7,9,17,20). CT scan and MRI demonstrate abnormal dilation of midline venous structure. CT scan was generally unable to provide detailed information regarding flow pattern (1,7). Duplex Doppler sonography may be diagnostic in patients in whom the vein of Galen has enlarged but this finding is nonspecific (17). Four vessel angiography is the main diagnostic method but is an invasive procedure (10,17,20).

In TCD investigation, main feeding arteries of the AVM's were identified by high flow velocity (Vm 132 cm/sec), low pulsatility indices (PI 0.46) (normal ranges 37±7.6 cm/sec, 0.83±0.24 respectively (8), and decreased CO2 reactivity. Blood flow velocities in basal cerebral arteries were correlated with angiographic findings. Flow velocity measurements permitted noninvasive diagnosis of AVM. Furthermore, the identification of individual feeding arteries permitted good definition of the anatomical localization of individual AVMs.

Flow velocity levels in feeding arteries are characteristically higher than in normal arteries (16). The low pulsatility in AVM feeders is due to the combined effect of the low peripheral resistance in the AVM and the pressure distributed along the high velocity inflow channels (15).

The straight sinus anomaly is rarely documented. No filling or narrowing of straight sinus was demonstrated in several studies (12,18,20). Minakawa et al. (12) reported nine cerebral AVMs associated with straight sinus anomaly. These AVMs occupied deep cerebral structures. They suggested that the absence of filling and narrowing of the straight sinus are possibly congenital anomalies. Viñuela et al. (18) have pointed out that these abnormalities were more than likely secondary changes due to flow phenomena with sinus occlusion and the secondary development of alternative routes of venous drainage. Although AVM of the vein of Galen associated with straight sinus anomaly is extremely rare (20), our case report demonstrated stenosis of the straight sinus and the presence of an accessory straight sinus.

Although the treatment of galenic AVM is controversial, endovascular technique is recommended in the treatment of type 4 galenic AVM and in neonate with cardiac failure, type 1 is treated surgically, types 2 and 3 are treated either surgically or with combination of surgery and endovascular technique (6,7,10,11,20).

Our patient was operated on through the posterior interhemispheric route as described by Yaşargil et al. (19,20). As confirmed by TCD investigation preoperatively, the lesion was supplied by both PCAs but main feeders were from the left PCA. The feeders were coagulated and cut with microscissors.

In assessing flow patterns after endovascular or surgical therapy, different diagnostic methods can be used such as cerebral angiography, CT, color Doppler imaging (10,17,20). The best method is cerebral angiography which is an invasive method.

In conclusion, TCD investigation can be used to determine the main feeders preoperatively and to evaluate the effect of therapy in the postoperative period. The procedure is noninvasive and can be performed easily and repeatedly. Although our experience with endovascular technique is limited, we believe that in the neonate especially in those with cardiac failure treated with endovascular technique complete postoperative angiography is unnecessary and the follow-up may easily be made by TCD investigation.

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