We report a rare variation of the left persistent primitive trigeminal artery with direct anastomosis with the ipsilateral posterior cerebral artery in a 53-year-old woman. She was sent to the hospital with visual deterioration and consciousness disturbance (Glasgow coma scale = E4V3M5). Marked hydrocephalus was found. An associated giant pseudoaneurysm was shown arising from the cisternal segment (C6) of the left internal carotid artery (ICA). She received a Hunterian ligation of the left ICA for treatment of the aneurysm in another hospital 5 years previously. The neurosurgeon thereafter removed and clipped the pseudoaneurysm to relieve its mass effect against the optic chiasm. She died of the complication of sepsis following extensive infarction of the territory of the left anterior cerebral artery (ACA) 6 weeks later. This case stresses the significance of careful initial full 4-vessel survey, balloon test occlusion with neurophysiologic monitoring before surgical ligation if fetal circulation is present, and alternative endovascular treatment of the pseudoaneurysm using the GDC (Guglielmi detachable coil) system.

Keyword: Persistent primitive trigeminal artery; Anomalous posterior cerebral artery; Pseudoaneurysm; Hunterian ligation; Guglielmi detachable coil (GDC)

The presence of persistent fetal circulation plays an important part in cerebral vascular intervention. There are mainly 4 types of embryonic carotid-basilar anastomoses, namely the trigeminal artery, the acoustic (otic) artery, the hypoglossal artery and the proatlantic intersegmental artery [1]. The persistent primitive trigeminal artery (PPTA) is the predominant type and is seen in 0.1% to 0.6% of cerebral angiograms. We report a variation of PPTA which directly anastomosed with the ipsilateral anomalous posterior cerebral artery (PCA). To our knowledge, this variation is rarely documented in the international literature.

The aneurysm often accompanies the persistent fetal circulation. It may be located at or near the origin of the persistent primitive arteries. It may also be situated far from the orifice of these primitive arteries and the relationship between the aneurysm and the persistent primitive arteries remains questionable. For a giant intracranial aneurysm, Hunterian ligation had been the preferred method. Due to rapid development of interventional neuroradiology, alternative endovascular treatment using the Guglielmi detachable coil (GDC) system has gradually taken the place of surgical clipping. Retrospectively, we also present this case to emphasize the significance of a careful initial imaging study and choice of therapeutic method to avoid a life-threatening situation.
CASE REPORT

A 53-year-old woman was sent to the hospital due to consciousness disturbance with Glasgow coma scale of 12 (E4V3M5). The right pupil light reflex was absent. Some underlying diseases included diabetes mellitus, acute myocardial infarction, upper gastrointestinal bleeding and cerebrovascular disease. She had received a Hunterian ligation of the left ICA for surgical treatment of a giant left ICA aneurysm in another hospital 5 years previously. Progressive visual deterioration after operation was reported by her family. The original imaging study was not available. A series of examinations were obtained, including computed tomography (CT), magnetic resonance (MR) imaging with contrast-enhanced magnetic resonance angiography (CE-MRA), and digital subtraction angiography (DSA).

Non-contrast CT initially showed a giant suprasellar aneurysm with marked hydrocephalus, but the vascularity was not well identified (Fig. 1). DSA via the left common carotid artery did not disclose the branches of the left ICA due to complete ligation. The right ICA and its branches also revealed no opacification of an aneurysmal lesion. However, the angiogram via the left vertebral artery (VA) clearly showed a spade-shaped aneurysm at the cisternal segment (C6) of the left ICA (Fig. 2). The distal portion of the left ICA on the ligated site was abnormally opacified. The supply artery was recognized as a left

Figure 1. Axial non-contrast CT shows a huge suprasellar inhomogeneous hyperdense mass, mimicking a calcified neoplasm. Marked hydrocephalus is noted. Evidence of left craniotomy is demonstrated.

Figure 2. DSA of the left VA. Left posterior oblique projection. A spade-shaped aneurysm (curved arrow) is found at the cisternal segment (C6) of the left ICA. The intracavernous segments of the left ICA are opacified. A tortuous engorged artery, namely the PPTA (arrow), originating from the presellar segment (C5) of the left ICA courses posteriorly and is continuous with the caudally displaced anomalous left posterior cerebral artery (PCA). ACA, anterior cerebral artery; MCA, middle cerebral artery.

Figure 3. Sagittal precontrast T1-weighted image (TR/TE/Excitation=450/12/2). The PPTA tortuously courses cephalad and posteriorly, and indents the dorsum sellae (arrows).
persistent primitive trigeminal artery (PPTA). It ran off the left ICA at the presellar segment (C5), indented the dorsum sellae (Fig. 3), and anastomosed with the anomalous left PCA (Fig. 4). The left PCA did not course normally and symmetrically with the contralateral side but was caudally displaced. The left posterior communicating artery (PCom) was not well depicted on either conventional or MR angiography (MRA) and was considered to be hypoplastic or agenetic, which was proven during surgery.

During surgery, a giant aneurysm was found inside an organized pseudoaneurysm which was grossly coated by dark hemosiderin. The axial T2-weighted images well demonstrated hemosiderin deposition (Fig. 5) due to repeated hemorrhage. After episodes of subarachnoid hemorrhage with thrombosis and organization, the residual patent lumen was the true aneurysm which was seen on both conventional and MR angiographic images. It was embedded within the giant pseudoaneurysm and was well shown on cross-sectional MR images (Fig. 6).

To relieve the mass effect of the giant pseudoaneurysm, the neurosurgeon removed and clipped it. Unfortunately, extensive infarction of the territory of the left ACA was noted on subsequent CT examination, complicated with pneumonia and sepsis which led to her death 6 weeks later.

**DISCUSSION**

The presence of fetal circulation in carotid-basilar anastomoses is of important clinical significance for intracranial surgical intervention and endovascular management. Though the PPTA is the predominant carotid-basilar anastomosis, its direct connection with the posterior cerebral artery is relatively rare according to the literature. We documented one case of this variation.

The PPTA was first shown angiographically by Sutton in 1950 [2]. It is seen in 0.1% to 0.6% of cerebral angiograms and it arises from the ICA between the presellar (C5) and juxtasellar (C4) segments. The other embryonic carotid-basilar anastomoses include the acoustic (otic) artery, the hypoglossal artery, and the proatlantic intersegmental artery. Usually the primitive arteries occlude and the PCom develops and becomes the predominant carotid-basilar communicating artery. The reason for the lack of closure versus reopening of the primitive arteries is not clear but some protective mechanism is assumed [3]. The PPTA may join the basilar artery between the anterior inferior cerebellar arteries.
artery (AICA) and superior cerebellar artery (SCA). In this condition, the PCom may be absent, and the proximal basilar artery to the junction may be hypoplastic. It was classified as type I by Saltzman [4]. It may also join the SCA directly without absence of the PCom and hypoplasia of the proximal basilar artery, establishing type II of Saltzman [4]. Different types of arterial variants and anomalies associated with the PPTA have been reported [5,6,7]. In our case, it fused with an anomalously coursed left PCA and the PCom was absent. Some might think it to be an entirely anomalous PCA per se.

This ICA pseudoaneurysm might be associated with the PPTA but this is questionable because it did not arise directly from the PPTA or near the PPTA’s origin from the ICA [8,9]. For surgical intervention, successful shrinkage of an ICA aneurysm which cannot be clipped depends on a significant decrease of ICA blood flow. In our case, total ligation of the left ICA cut off the carotid supply but the basilar supply remained. Obviously this treatment failed and the symptoms worsened. To reverse this condition, the collateral circulation, more commonly the PPTA, should be investigated and ligated simultaneously. Alternatively, ligation of the ICA distal to the conjoint site of the ICA and the PPTA may also be effective. Performing an extracranial-intracranial (EC-IC) bypass is another preoperative consideration, to avoid insufficient blood supply to the distal branches of the main cerebral arteries [10,11]. Preoperative balloon test occlusion with neurophysiologic monitoring should be used to evaluate hemodynamic changes. In the absence of significant hemodynamic change, Hunterian ligation had been the preferred method for treatment of a giant intracranial aneurysm. Follow-up angiography is important because recanalization after Hunterian ligation may occur [12].

Non-surgical therapy for aneurysms has developed rapidly. Over the past decades, interventional neuroradiologists have tried balloons, glues, and various coils to embolize aneurysmal sacs. The introduction of the GDC system has revolutionized endovascular treatment. It was designed in the early 1990s and received FDA approval in 1995. It is available in numerous sizes and shapes, and has soft and atraumatic features. Repositioning of the GDC system in an aneurysmal sac is possible and many placements and retrievals can be done until a satisfactory position is obtained. Its effect is far more obvious in ruptured aneurysms than in unruptured ones as there is a rehemorrhage rate of approximately 1% to 2% at 6 months [13].

With our case, it was difficult to initially choose whether to use surgical clipping, endovascular treatment or both because of the patient’s deteriorated visual acuity. In addition to reporting the variations in the PPTA and anomalous posterior cerebral artery, we would also like to emphasize a careful full 4-vessel angiographic survey initially to discover the location of the aneurysm and possible presence of fetal circulation if MRA or computed tomography angiography (CTA) is not sufficient.

Endovascular therapy using the GDC system might have been more effective in this patient before her visual acuity change and she might have survived longer because the pseudoaneurysm might not have developed this huge to produce significant mass effect.

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變異的存續原三叉動脈與巨型僞內頸動脈瘤

林威辰¹  聶國倫²  謝文郁³  劉金昌¹

高雄醫學大學附設中和紀念醫院 放射線科¹
嘉義華慈醫院 放射診斷科²  神經外科³

存續性原三叉動脈雖是胚胎腦部血液循環中較常見的一種，但直接與後大腦動脈相連則是較為少見的變異，我們曾報了一例有此變異的53歲女性。此外，該患者同時發現有一相當巨大的僞內頸動脈瘤位於內頸動脈的第六節（池節）上，且曾於五年前在他院接受過漢他氏結紮手術。此時因此瘤過大造成視交叉壓迫，外科醫師遂施行移除手術並緊以外科夾；其後因發生廣泛性左前大腦動脈供應區梗塞，並合併敗血症，於十週後導致死亡。這個不幸的經驗強調了最初四條腦血管攝影評估的重要性，且若發現存續的胚胎血液循環要以外科方法結紮時，應先做氣球測試性堵塞及神經生理監視，以觀察患者是否合適結紮。如不使用傳統外科夾除手術處置，於今日日趨成熟的高尼米可分離式線圈是另一替代性的血管內治療方法。

關鍵詞：存續性原三叉動脈；異常的後大腦動脈；僞動脈瘤；漢他氏結紮；高尼米線圈