

Carotid Agenesis with Intercavernous Anastomosis

İnterkavernöz Anastomoz ile Karotid Agenesisi

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ABSTRACT

Agenesis of internal carotid artery (ICA) is rare. Association of agenesis with abnormal arterial communication between the cavernous segment of the carotid arteries is extremely rare. We present a case of a middle-aged woman who presented with sudden onset severe holocranial headache. She was investigated and diagnosed to have sub-arachnoid hemorrhage on CT scan. Digital Subtraction Angiogram revealed absent right internal carotid artery. The right middle cerebral artery received blood supply from an incidentally detected intercavernous anastomosis from the left internal carotid artery. No aneurysm, arteriovenous malformation or any other vascular abnormality was detected. The patient was managed conservatively. She has been asymptomatic for five years on follow up.

KEYWORDS: Agenesis, Internal carotid artery, Cavernous sinus, Subarachnoid hemorrhage

ÖZ

İnternal karotid arter (İKA) agenezisi nadirdir. Agenezinin karotid arterlerin kavernöz segmentleri arasında anormal arteriyel iletişim ile ilişkili olması çok nadirdir. Ani başlangıçlı şiddetli holokraniyal baş ağrısı ile gelen orta yaşlı bir kadın sunuyoruz. İncelemeler sonucunda BT tarama ile subaraknoid kanama tanısı konmuştur. Dijital Subtraksiyon Anjiyogramı sağ internal karotid arterin bulunmadığını göstermiştir. Sağ orta serebral arterin sol internal karotid arterden tesadüfi olarak saptanan bir intrakavernöz anastomoz yoluyla kan aldığı görülmüştür. Herhangi bir anevrizma, arteriyovenöz malformasyon veya başka vasküler anomali saptanmamıştır. Hasta konservatif olarak takip edilmiştir. Beş yıllık takipte asemptomatiktir.

ANAHTAR SÖZCÜKLER: Agenez, İnternal karotid arter, Kavernöz sinüs, Subaraknoid kanama

INTRODUCTION

Agenesis of the internal carotid artery (ICA) is a rare congenital developmental anomaly. Agenesis of the ICA is usually asymptomatic and is often detected as an incidental finding after a cerebrovascular event such as subarachnoid hemorrhage (SAH) or cerebral infarct (4). In such cases of absent ICA, the contralateral ICA or basilar artery (BA) supplies the middle cerebral artery (MCA) and anterior cerebral artery (ACA). Approximately 17 cases of an unusual intercavernous anastomotic vessel connecting the ICA have been reported in the literature (1). The present case is being reported because of the rarity of this type of lesion.

CASE REPORT

A 52-year-old lady presented with sudden onset severe holocranial headache of two-day duration without any loss of any consciousness or vomiting. On examination, the patient was conscious and there was minimal weakness of the right lower limb. BP was 130/70 mm Hg. Computed tomography (CT) scan of the head showed subarachnoid hemorrhage in the interhemispheric and sylvian fissure (Figure 1A). Skull base CT scan demonstrated complete absence of the right carotid canal (Figure 1B). Digital subtraction angiography (DSA) showed absence of the right ICA in the neck and the

petrous portion (Figure 2A). The middle cerebral artery on the right was filled from the contralateral ICA through an intercavernous connection (Figure 2B). No aneurysm was seen on DSA. MR angiography also showed absence of the right ICA. The patient has been asymptomatic for the last 5 yrs.

DISCUSSION

Internal carotid agenesis is rare congenital anomaly. Certain mechanical and hemodynamic stresses on the embryo like exaggerated folding of the embryo towards one side and constriction by amniotic bands have been postulated as cause for unilateral agenesis (6).

Lie (2) described six pathways of collateral circulation in association with absence of ICA and our case belongs to type D in which there is intercavernous communication to the ipsilateral siphon from the contralateral cavernous ICA. The intercavernous collateral may be located posterior to the clivus or run above through or in the floor of sella turcica. On imaging, the transsellar variety of the vessel may mimic a pituitary microadenoma. In this type of intercavernous communication there is associated hypoplasia or aplasia of the ipsilateral A1 segment of the anterior cerebral artery with the patent anterior communicating artery supplying the

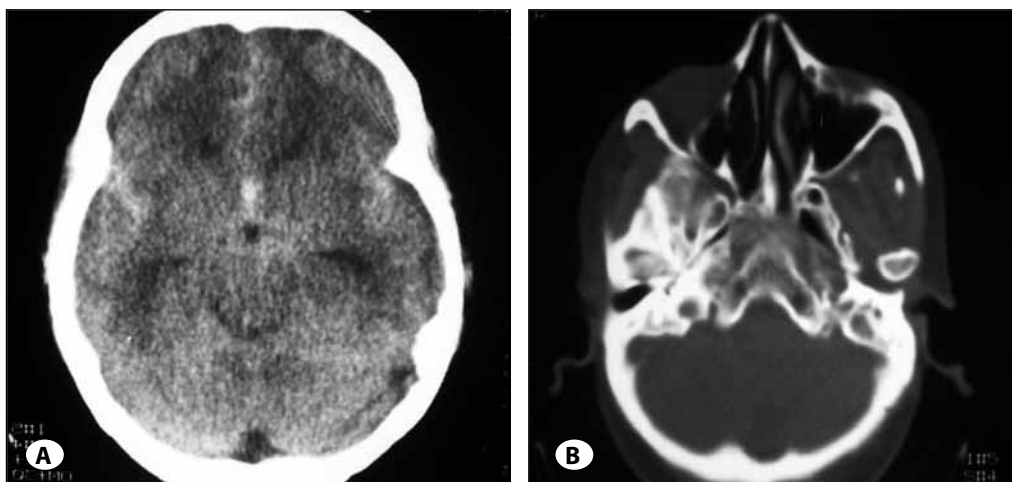


Figure 1: A) Axial CT scan brain window showing subarachnoid hemorrhage in the inter hemispheric and sylvian fissure. B) Axial CT scan bone window showing the absence of the right carotid canal.



Figure 2: A) Right common carotid angiogram showing right external carotid branches and absence of internal carotid artery. B) Left common carotid angiogram showing anomalous collateral vessel connecting the cavernous portion of the right and left internal carotid arteries further supplying the right MCA. The right ACA is filling from a patent anterior communicating artery and the right A1 segment is absent.

ipsilateral distal anterior cerebral artery territory. The majority of the ipsilateral middle cerebral artery supply is derived from the intercavernous communication isolating it from the alternate collateral flow.

Male preponderance has been reported in the literature for these patients and there was equal involvement of the right and left side (5). Four of these 18 patients including one of ours had an aneurysm (22.2%), which is much higher than the 2-4% natural incidence of intracranial aneurysm. Our patient presented with subarachnoid hemorrhage but no aneurysm could be demonstrated on DSA. She has been asymptomatic for the last five years. The high incidence of aneurysm in carotid

agenesis is ascribed to the altered hemodynamics of blood flow through collateral channels (1), hence these patients should be followed up both clinically and radiologically for the development of an aneurysm.

According to Lasjaunias & Vazquez (3), the internal carotid artery is formed by the seven distinct segments. Each segment is independent and may show agenesis. Agenesis of one segment leads to regression of all segments proximal to the anomaly. In our case there was agenesis of the third segment resulting in the filling from the contralateral internal carotid artery via the primitive maxillary artery.

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