

## A RARE ENCOUNTER IN PRIMARY CARE: INTERNAL CAROTID ARTERY AGENESIS-CASE REPORT

*Birinci Basamakta Nadir Görülen bir Olgu: İnternal Karotis Agenezisi Olgu Sunumu*

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### ABSTRACT

Headache is a quite common presenting complaint in primary care. Detailed examination and imaging are essential for differential diagnosis, as they can reveal various underlying pathologies. In this article, we present a case of internal carotid artery (ICA) agenesis which is detected during the investigation for headache. A patient presented to the family medicine clinic with a two-month history of persistent headache. The general examination did not reveal any pathology or neurological signs. Due to a family history of aneurysms, the patient was referred to the hospital for imaging. Magnetic Resonance Angiography showed unilateral ICA agenesis in the patient, and the patient was placed under follow-up to prevent accompanying cerebrovascular diseases. ICA agenesis often follows a symptomatic course but occasionally manifests with neurological symptoms. When this condition is detected, taking vascular protective measures and recommending lifestyle changes can be long-term steps to prevent pathologies such as aneurysm rupture or ischemia for the patient.

**Keywords:** Internal carotid artery, headache, agenesis, aplasia

### ÖZ

Baş ağrısı, birinci basamakta oldukça sık karşılaşılan bir şikayettir. Detaylı muayene ve görüntüleme, altta yatan çeşitli patolojileri ortaya çıkarabilme bakımından ayırıcı tanıda esastır. Bu makalede, baş ağrısının araştırılması sırasında tespit edilen bir internal karotis arter agenezisi olgusunu sunuyoruz. Hasta, 2 aydır süregelen baş ağrısı şikayetiyle aile hekimliği kliniğine başvurdu. Genel muayenede, herhangi bir patoloji veya nörolojik belirti tespit edilmedi. Ailesinde anevrizma öyküsü olması nedeniyle hasta, görüntüleme için hastaneye yönlendirildi. Manyetik Rezonans Anjiyografi, hastada tek taraflı internal karotis agenezisini gösterdi ve hasta, bu duruma eşlik edebilen serebrovasküler hastalıkları önlemek için takip altına alındı. İnternal karotis agenezisi sıklıkla semptomatik bir seyir izlese de nadiren nörolojik belirtiler eşliğinde görülmektedir. Bu durum tespit edildiğinde, vasküler koruyucu önlemler alınması ve yaşam tarzı değişiklikleri önerilmesi hastada ilerleyen zamanda anevrizma rüptürü veya iskemi ortaya çıkmasını önlemeyen uzun dönem koruyucu adımlardır.

**Anahtar Kelimeler:** İnternal karotis arter, baş ağrısı, agenezi, aplazi



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## INTRODUCTION

The internal carotid artery (ICA) plays a pivotal role in delivering oxygen and nutrients to the brain, making any anomalies or pathologies in this vessel of paramount clinical importance (1). One such rare anomaly is the agenesis of the internal carotid artery, a condition characterized by the complete absence of the vessel. Although cases of ICA agenesis are relatively infrequent, their clinical implications can be profound and pose significant challenges to diagnosis and management.

This case report aims to shed light on a case of ICA agenesis, to provide a comprehensive overview of the patient's clinical presentation, diagnostic workup, and therapeutic considerations. Drawing from our clinical experience and the available literature, we will discuss the intricate vascular anatomy, potential etiologies, and clinical implications of this rare condition.

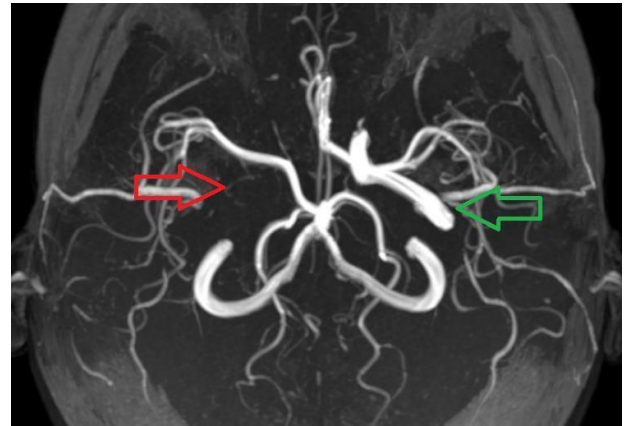
## CASE REPORT

A 26-year-old female patient presented to the family medicine clinic with a complaint of recurrent headaches occurring more than three days a week for approximately 2 months. The patient described the headache as a constant heaviness throughout her head. When asked to rate the pain on a scale from 0 to 10 (with 10 being unbearable pain and 0 being no pain), the patient rated it as 6. The headache did not interfere with her daily activities, and it did not wake her up from sleep. The intensity of the pain was not affected by light or sound. The patient mentioned that she felt better when lying down but experienced a slight increase in pain, her especially during physical exertion while standing. There were no accompanying symptoms such as nausea, numbness, or tingling. The patient had not experienced any significant, continuous headaches until two months ago, and she did not have any underlying medical conditions.

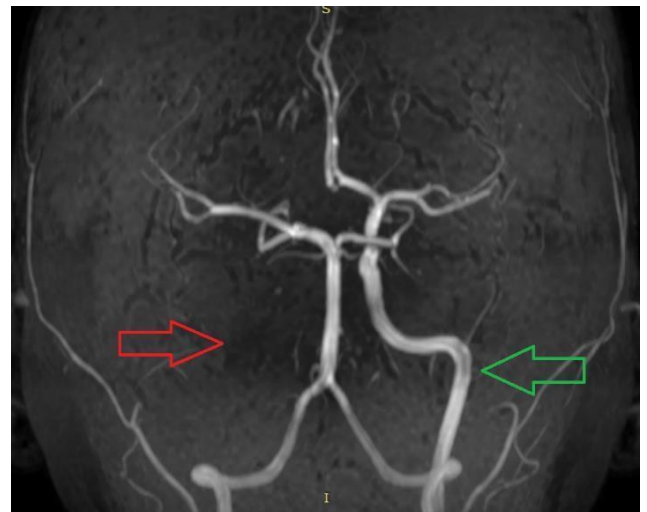
The patient, who worked as an accountant in a company, had routine blood tests done about 3 weeks ago during a corporate health screening, which did not reveal any abnormal values. Her hemoglobin was 14.2 mg/dL, and her hematocrit was 37.2%. Her blood pressure was measured at 118/74 mmHg. A neurological examination did not show any pathological findings, and there were no signs of meningeal irritation. The patient had regular menstrual cycles, and there were no significant life events affecting her psychological well-being in the past 2 months. She did not follow any specific dietary patterns or use any supplements. An ophthalmological examination conducted within the last 6 months did not detect any visual impairment. There were no chronic illnesses among the patient's first-degree relatives, but her uncle had passed away at the age of 48 due to a cerebral aneurysm.

Despite a detailed physical examination that did not reveal any pathological findings, considering the possibility of cerebrovascular pathology and the need to rule out intracranial causes, the patient was referred to the neurosurgery clinic for further evaluation.

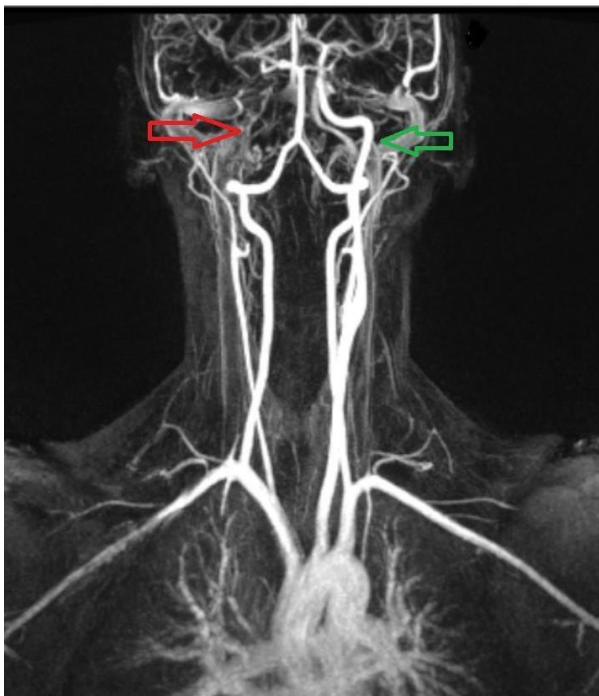
After being examined by a neurosurgeon, the patient underwent a magnetic resonance angiography (MRA), and she returned with the results a week later. The MRA revealed the absence of the patient's right ICA (Figures 1,2 and 3). While the headache was thought to be related to this condition, there was no urgent need for intervention. Nevertheless, due to the possibility of an associated aneurysm or ischemia, the patient was placed under surveillance for cerebrovascular risks. She was informed about blood pressure control and lifestyle changes, including dietary modifications to prevent hyperlipidemia.



**Figure 1:** Cerebral axial MRA image: The left ICA is visible indicated by the green arrow, while on the right side, the artery is not observed (red arrow).



**Figure 2:** Cerebral coronal MRA image: The left ICA is visible indicated by the green arrow, while on the right side, the artery is not observed (red arrow).



**Figure 3.** Cervicocranial MRA image: The left ICA is visible indicated by the green arrow, while on the right side, the artery is not observed (red arrow).

## DISCUSSION

The congenital absence of the ICA, which can manifest as agenesis, aplasia, or hypoplasia, is a rare condition occurring in less than 0.01% of the general population (2). However, a more recent retrospective study found this rate to be 0.13% (3). While it is mostly unilateral, the absence of the left ICA is reported to be 3 times more frequent by some authors and 1.5 times more frequent by others compared to the right side (4,5). In our case, the absence of the right ICA is observed.

Although the terms agenesis and hypoplasia are sometimes used interchangeably, their definitions differ anatomically and embryologically. In agenesis, both the ICA and the carotid canal are absent, while in aplasia and hypoplasia, a portion of the vessel with a widened proximal segment is visible, and the canal is present (4,6).

The majority of cases of ICA agenesis remain asymptomatic due to the development of extensive collateral circulation, which prevents cerebral ischemia (7). However, in rare instances, the diagnosis can be made through the investigation of symptoms such as tinnitus, headache, or blurred vision (8). In this case, the patient's complaint leading to the clinic visit was a headache.

It is reported that various cerebrovascular anomalies may accompany ICA agenesis. Particularly, the incidence of aneurysms is much higher in individuals with ICA agenesis compared to the normal population (25-43% vs. 2-4% respectively) (9-11). Severe presentations -such as ischemia or subarachnoid

hemorrhage- are believed to be related to the rupture of accompanying aneurysms (8,12). The underlying mechanisms for aneurysms occurring alongside ICA agenesis are thought to involve hemodynamic disturbances or embryonic developmental pathologies (13). While no accompanying aneurysm was detected in this case, the patient's family history of aneurysm rupture raises the possibility of a genetic predisposition. Family case series have been reported in both ICA agenesis and aneurysm cases, suggesting a genetic basis (14,15). However, no specific genetic cause has been identified in previous studies.

Despite its rarity, the significance of internal carotid artery agenesis lies in its potential to cause various neurological symptoms and the diagnostic dilemmas it presents. Through this case report, we hope to contribute to the growing body of knowledge surrounding this condition and foster a deeper understanding of its management and tailored risk minimization strategies. The authors declare no conflict of interest. This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

Informed consent has been obtained from the participants for the study.

*Researchers' Contribution Rate Statement:*  
Concept/Design: BUK; Analysis/Interpretation: OK;  
Data Collection: OK; Writer: BUK; Critical Review:  
OK; Approver: OK

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