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Unilateral cervical and petrosal segment agenesis of the internal carotid artery with rete mirabile

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ABSTRACT

The carotid rete mirabile (RM) is a physiological network between the internal and external carotid arterial systems. In this paper, an extremely rare case is presented, in which internal–external carotid artery anastomoses and a dilated ascending pharyngeal artery, due to unilateral agenesis of the cervical and petrous segments of the internal carotid artery (ICA), is presented.

1. Introduction

Collateral webs are very rare in humans unless aplasia, hypoplasia, or stenosis of the native vessels occurs. These vessels can be crucial to maintaining antegrade blood flow in the vascular territory of the aplastic or hypoplastic artery. Carotid RM is defined as the segmental agenesis of the epidural portion of the ICA, with a compensatory arterial network supplied by the external carotid artery (ECA) branches—frequently, the branches of the maxillary artery and ascending pharyngeal artery [1]. Clinically, patients with carotid RM can manifest with vertigo, headache, loss of balance, subarachnoid bleeding related to an aneurysm, carotid fistula, pseudoxanthoma elasticum, or stroke at a young age; however, they can also be found incidentally during cross-sectional imaging studies [2]. Knowledge of the complex anatomy, functions, clinical significance, and embryological development of carotid RM is critical to its treatment. While a few cases of bilateral RM have been reported [2–6], unilateral RM is extremely rare. Herein, the complicated anatomy and potential pitfalls of a unilateral carotid RM case are described.

2. Case

A 33-year-old woman was referred to an ear, nose, and throat clinic because she had been experiencing pain in her left ear, reflecting down to her chin, for three months. Her physical examination was normal. Using Doppler ultrasonography (US), no pathology was seen in the lumen of the left common carotid artery (CCA), ICA, or ECA; however, a high resistive index (RI) was detected (0.80) in the ICA (Fig. 1). Computed tomography (CT) angiography showed that the left ICA was dysplastic, and the carotid canal was hypoplastic (Fig. 2). Magnetic resonance angiography (Fig. 3) revealed an extremely dilated dominant branch (the superior pharyngeal artery) of the ascending pharyngeal artery, which was forming the C3 (lacerum) segment of the ICA after entering into the skull base through the foramen lacerum.

Four system of digital subtraction angiography and 3D rotational angiography (Figs. 4 and 5) revealed that the C1 and C2 segments of the left ICA were absent and the distal part of it (C3–7) had antegrade blood flow resulting from collateral of the internal maxillary and ascending pharyngeal arteries. An extremely dilated ascending pharyngeal artery was shown as entering into the skull base via the foramen lacerum by its dominant superior pharyngeal branch; then, anastomosis with the meningohypophyseal truncus was detected (Figs. 6, 7).

The collateral web was suggested to be an RM according to the patient's symptoms, the absence of atheroma, and the angiography data. The clinical symptoms of the patient were spontaneously resolved, and no medical or surgical therapy was required.

3. Discussion

Cerebral collateral circulation can be categorized into three types: cerebral collateral circulation through the circle of Willis, through the persistent fetal circulation, or through the skull base arteries exiting from the ECA (called RM) [7,8]. Nevertheless, while RM is unusual in humans, it is frequently seen in vertebrates; their Willis circle is defective in cases where it is compensated by maxillary and infrequently vertebral arteries. In some mammals, during the embryologic development of RM, ICA secondarily undergoes atrophy, and distal ICA flow is then reestablished by embryologic arteries, such as the ascending

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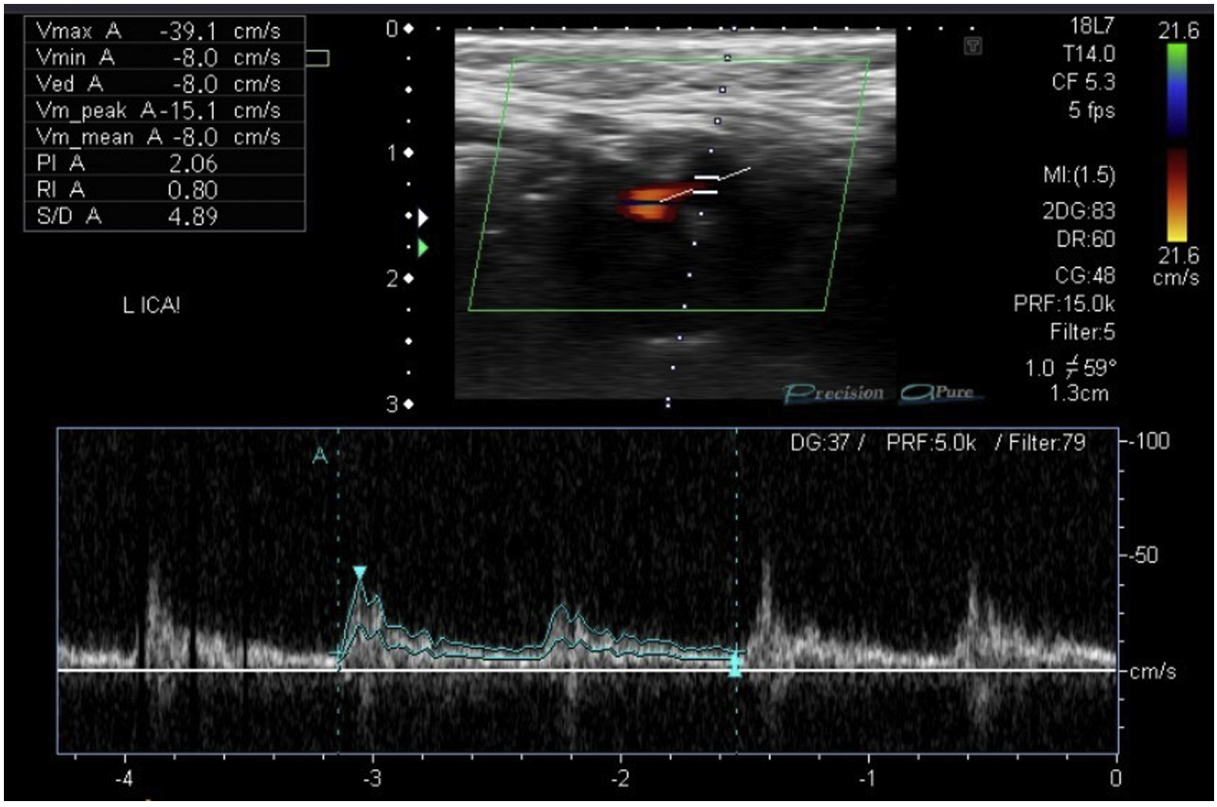


Fig. 1. Doppler ultrasonography showed no luminal pathology in the left CCA, ICA, and ECA but increased resistivity index (RI = 0.80) was detected.

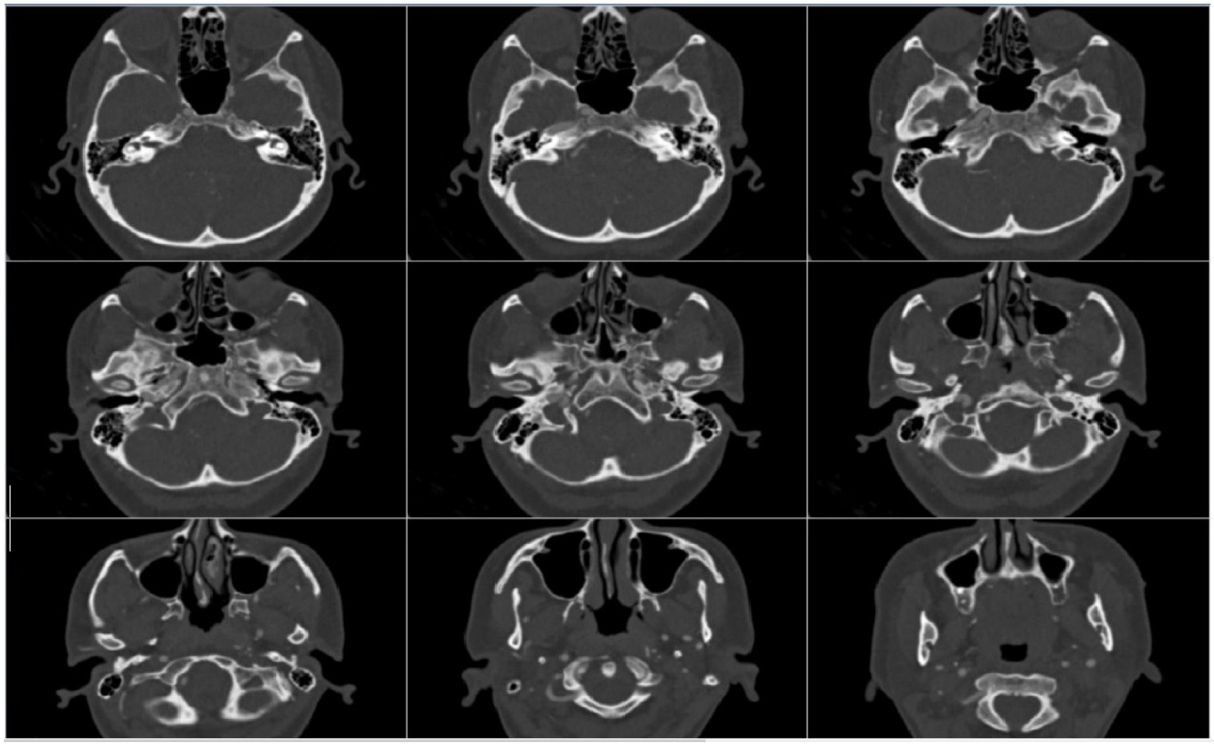


Fig. 2. Cranial CT showed that the left carotid canal was hypoplastic.

pharyngeal, basilar, trigeminal, primitive maxillary, and internal maxillary arteries [1]. The secondary atrophy of ICA is physiological in pigs, cows, sheep, and goats, to maintain the arterial supply of the Willis circle. It has been suggested that this frequent adaptation regulates intracerebral blood pressure and flow, to facilitate thermoregulation [1,2].

Normally, the carotid RM is not present in any human developmental state. It has been postulated that the absence of ICA occurs through regression after its development (in the fetal or perinatal period) and that the carotid RM develops earlier, as a secondary

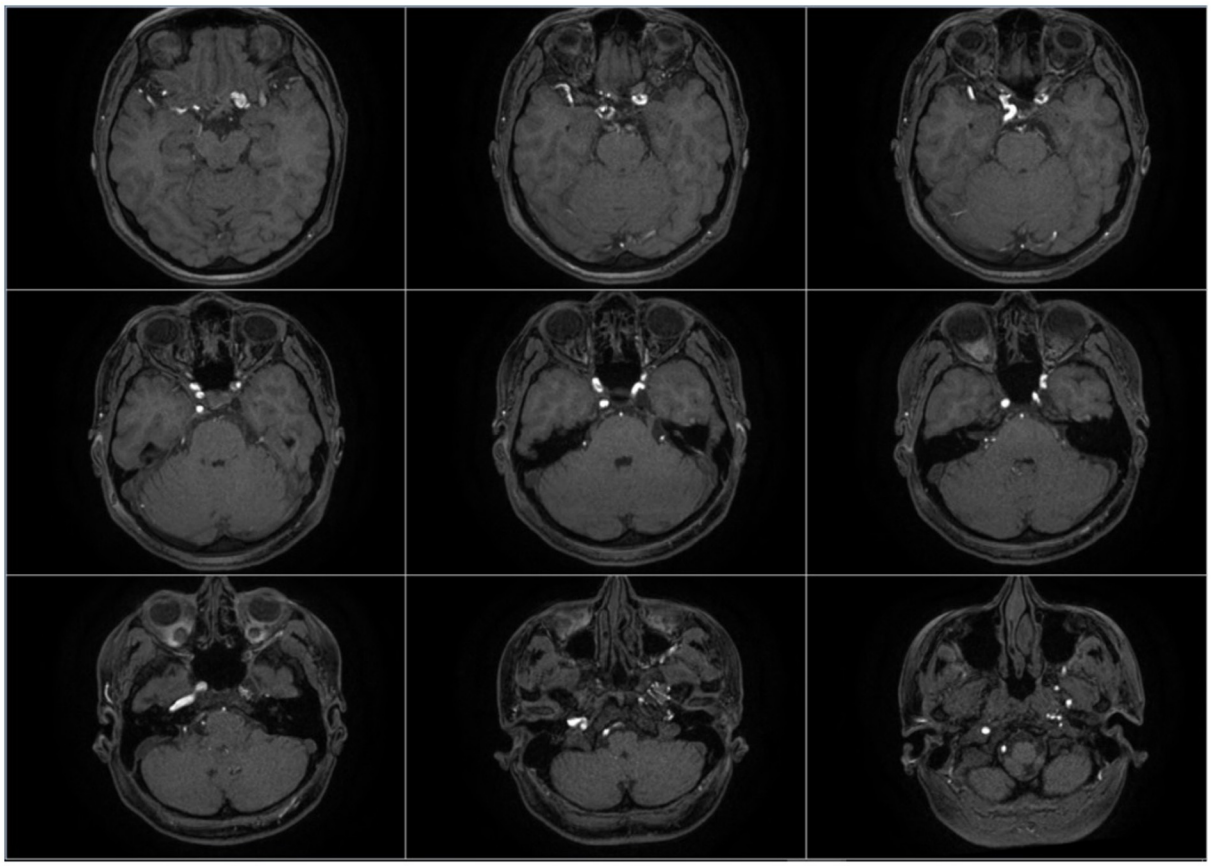
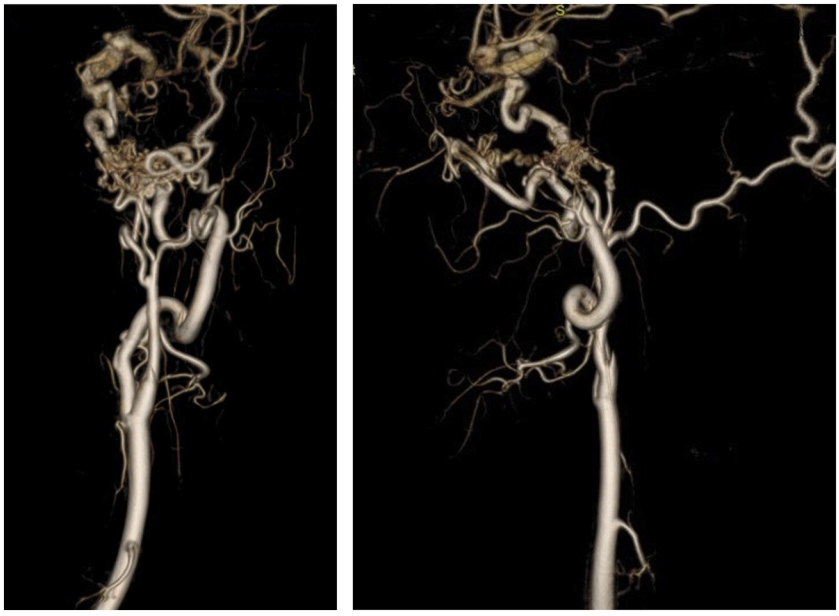


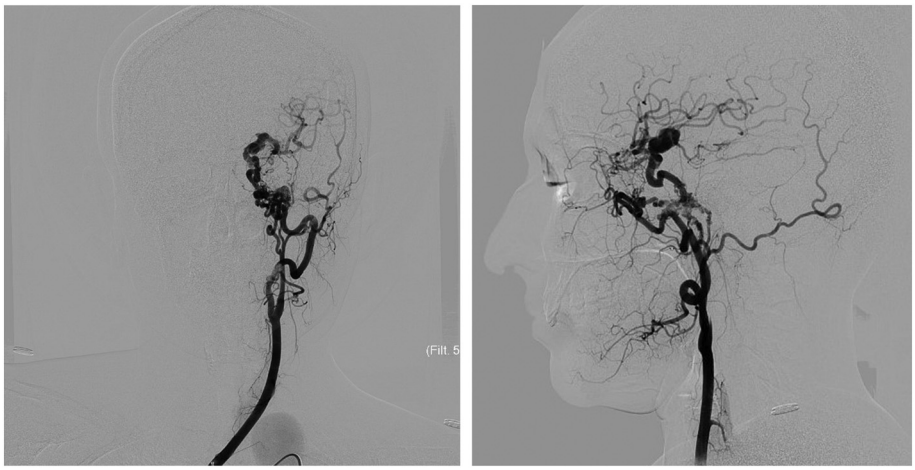
Fig. 3. MR Angiography. Agenesis of the cervical and petrous segments of the left ICA and numerous small vessels forming the lacerum segment of the ICA is seen. Additionally wide superior pharyngeal branch of the ascending pharyngeal artery forming the ICA after entering into the skull base via foramen lacerum.



Figs. 4, 5. The view of the left CCA angiography, anteroposterior and lateral 3D virtual angiogram. Agenesis of the cervical and petrous segment of the ICA and its distal reconstruction at the lacerum segment.

collateral pathway when the embryonic primary trigeminal artery and hypoglossal artery have already regressed [2,5]. The hypoplastic carotid canal seen on CT scans supports this theory because the presence and pulsation of the ICA precursor is required for the development and shaping of the carotid canal at 5–6 weeks of gestation [2].

Carotid RM has the following characteristic features on angiogram: i) hypoplasia of the ICA beginning from the carotid bifurcation; ii) a rich arterial plexus supplied mostly by the branches of the ECA, especially those of the internal maxillary artery; iii) the supraclinoid ICA is not hypoplastic and is supplied by the arterial network and dilated



Figs. 6, 7. Left CCA angiographies, lateral and AP view. At the level of lacerum segment, the left ICA was distally reconstructed by the collaterals of the internal maxillary artery and meningohypophyseal truncus entering into the skull base via hypoglossal canal anastomosing with dilated clival branches of the ascending pharyngeal artery.

ophthalmic artery; iv) no other abnormal vessels, as seen in moyamoya, are found in the intradural circulation; 5) bilateral lesions [2,5]. Our case is therefore compatible with an RM diagnosis; however, in our case, unilaterality was observed. Additionally, the ipsilateral hypoplastic carotid canal seen on the CT scan is another radiologic characteristic of RM.

Bilateral epidural carotid artery RM cases are seen relatively frequently, but unilateral vertebral RM (i.e., at the junction of extra and intradural parts) and ophthalmic RM are extremely rare variations. The reported symptoms of unilateral RM include vertigo, loss of balance, subarachnoid bleeding related to an aneurysm, carotid fistula, pseudoxanthoma elasticum, and stroke [9]. The most clinically important condition is hemorrhagic and ischemic events at a relatively young age, which is relatively common in carotid and vertebral RM [2]. Persistent collateral circulation has been shown to be a potential source of emboli into the retinal or intracranial territories [10]; Lu et al. [5] reported a case of bilateral carotid and vertebral RM presenting with a headache in the left temporal area. Our case presented with left ear pain reflecting the in maxilla for three months, which could be explained by a steal from the maxillary artery or branches of the ascending pharyngeal artery (i.e., the sphenopalatine, descending palatine branches, and inferior tympanic branch of the caroticotympanic branch) [10]; nevertheless, it is uncertain why the symptom was transient. We encountered only one case of unilateral carotid RM [1]; this case was that of a 21-year-old woman who presented with ipsilateral facial paresthesia and dizziness; the symptoms resolved spontaneously without surgical or medical treatment.

RM cases should be differentiated between acquired compensation or vasa vasorum or “pseudo-rete.” RM is thought to occur during embryologic development; in contrast, vasa vasorum is a collateral web emerging due to occlusions or stenoses in later life. Vasa vasorum supplies microcirculation in 1/3 of the adventitia and the media of large and medium-size arteries, including the carotid artery. Vasa vasorum is not normally seen radiologically, but in patients with atherosclerosis, it can be visible because of neovascularization and neoangiogenesis. In this situation, vasa vasorum could be visualized on carotid Doppler US; nevertheless, it can also be misinterpreted as recanalization. Selective angiography must be done to verify the diagnosis. Vasa vasorum is exiting from the superior thyroideal and the ascending pharyngeal, forming a dense web in the atheroma [2,5]. It is mainly seen in proximal vessels, but there is a low possibility that it will occur in distal cranial arteries, such as the middle and anterior cerebral arteries. Rarely, in cases of complete occlusion of the ICA because of surgery or atherosclerosis, vasa vasorum can be large enough to maintain antegrade flow. Risk factors of atherosclerosis must therefore be considered in the management of these patients. The enlargement of atherosclerotic plaques is related to the steady development of vasa vasorum [11].

It is well known that the intra-extracranial collateral pathways may enlarge when ICA occludes. In such cases, the ascending pharyngeal to the cavernous ICA collateral may enlarge to a size sufficient to be confused with a hypoplastic or atherosclerotic ICA. Differentiation is crucial if surgical or interventional attempts to increase cerebral flow are considered. Lateral projections have little value in differentiating between narrowed ICA and ascending pharyngeal artery, but there are distinct anatomic differences within antero-posterior projections: The petrous segment of the ICA has three distinct segments, the vertical, genu, and horizontal segments. The lateral margin of the genu approaches the hypotympanum of the middle ear. The ascending pharyngeal collateral does not have distinct segments, and, most importantly, its lateral margin never approaches the hypotympanic area. In contrast, the course of the ascending pharyngeal artery is closely parallel to the ICA, except that the former is more medially located as it approaches the skull base. It gives off an inferior tympanic branch, terminating in the preclival nasopharynx; its distal branches anastomose in the nasopharynx with the branches of the ICA, including the vidian artery and the arteries that pass through the foramen lacerum and foramen ovale, which are usually derived from the posterior division of the ICA to the inferior cavernous sinus [12].

Through the common use of noninvasive radiologic diagnostic methods, anatomical variances are more frequently encountered. Although RM is an extremely rare abnormality, it can be found incidentally, or it can be the cause of a stroke in patients of a younger age. Embolization of the ECA branches is currently clinically important for the transarterial endovascular treatment of dural arteriovenous fistulas, traumas, and epistaxis and for the preoperative embolization of head and neck tumors before surgery to prevent surgical hemorrhage. The possible anastomotic pathways between the internal and external carotid systems must be well recognized to prevent major complications, such as embolic stroke during embolization procedures [13].

Digital subtraction angiography (DSA) is the gold standard imaging method to visualize the vascular anatomy with high spatial and temporal resolution. But DSA is expensive, time consuming; has vascular injury and nephrotoxic contrast medium complications and exposes ionizing radiation. Clinically CT and MR angiography has widely replaced the need of DSA imaging because of improvement of imaging technology and software. Bolus tracking techniques and high iodine delivery protocols enabled increased vascular enhancement in CTA; nevertheless CTA causes high radiation exposure, requires nephrotoxic contrast medium. Beam hardening and motion artifacts may degrade image quality. MRI and MRA offers the best diagnostic capacity to demonstrate soft-tissue components and relationships with nearby structures. Conventional non-contrast MRA techniques (such as time-of-flight and phase-contrast MRA), as well as contrast-enhanced MRA,

may allow vascular imaging but they have flow artifacts, they produce static images, and cannot detect small vessels; they are associated with early venous filling and overlapping with arteries. Time resolved imaging of contrast kinetics (TRICKS) is a new technique that offers better temporal and spatial resolution compared to contrast-enhanced MRA technique. This sequence provides dynamic images during the arterial, capillary and venous phases with 3-dimensional volumes during the passage of gadolinium. Vascular anatomy can be visualized similar to DSA with less motion artifacts. Its major advantage is significant reduction in contrast dose to prevent nephrotoxicity in renal insufficiency patients [14,15].

On color Doppler USG, the dilated common pharyngo-occipital truncus can be confused with the C1 segment of the ICA. An increase in the RI without atherosclerotic changes within the ICA lumen being considered can be a suspicious finding, indicating an abnormal connection between the internal and external carotid arterial systems. As far as we know, this finding has not been presented in English-written literature on the subject.

To sum up, in this study an extremely rare case of unilateral ICA agenesis and secondary anastomosis between the ICA and ECA, along with RM, is illustrated. Additionally, to the best of our knowledge, it is the first case of a dilated ascending pharyngeal artery with physiological angiogenesis of its collaterals for the compensation of unilateral ICA agenesis.

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