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Clinical Article

Segmental unfused basilar artery with kissing aneurysms: report of three cases and literature review

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Summary

Basilar artery ''fenestration'' is the result of a failed fusion of the bilateral longitudinal neural arteries and can be associated with a saccular aneurysm, which typically arises at the proximal juncture of the unfused segment. ''Kissing'' aneurysms at this site, i.e. two aneurysms arising from the proximal junction of the unfused segment of the basilar artery pointing anteriorly and posteriorly are reported to be exceedingly rare. We present three patients with this rare condition, all of them being treated by endovascular techniques.

Keywords: Basilar artery; kissing aneurysms; fenestration; duplication; unfused segment; anatomic variation.

Introduction

An arterial ''fenestration'' is a condition that results from a segmental non-fusion of two paired embryological vessels that fail to fuse during development [1, 2]. There is controversy regarding the site of the intracranial vascular system in which fenestration is most frequent. Some authors argue that it is at the vertebral arteries, followed by the basilar artery [3]. However, other series show that the basilar artery is the most common site of vessel fenestration, followed by the vertebral and middle cerebral arteries [4].

Basilar artery fenestrations have a frequency less than 6% in postmortem reviews and less than 1% in angiographic studies [1, 5–7]. Proximal basilar artery fenestrations (i.e. close to the vertebro-basilar junction) are more common than distal fenestrations [4, 8, 9]. The incidence of an associated aneurysm at the fenestration site is 3% in general, and up to 7% at the basilar artery [10]. Five patients with ''mirror image'' or ''kissing'' aneurysms at an unfused basilar artery, i.e. fenestrations that harbour two distinct aneurysms at the same site have been reported previously [4, 8, 11–13]. In the present publication, we describe three additional patients with this rare condition.

Case reports

Case 1

A sixty year old man presented with headache, transient loss of consciousness and left inferior limb motor deficit. Cranial axial tomography demonstrated subarachnoid hemorrhage (SAH). Digital angiography revealed a basilar artery fenestration associated with two kissing aneurysms, one posterior to the basilar artery measuring 8 mm and one 2 mm in size pointing anteriorly. There was also a 2 mm size left middle cerebral artery aneurysm. The 8 mm basilar aneurysm was probably the source of the hemorrhage and it was successfully embolised with coils (Fig. 1). Following this intervention, the patient had no neurological deficit initially, but during hospitalisation he developed dysarthria and somnolence

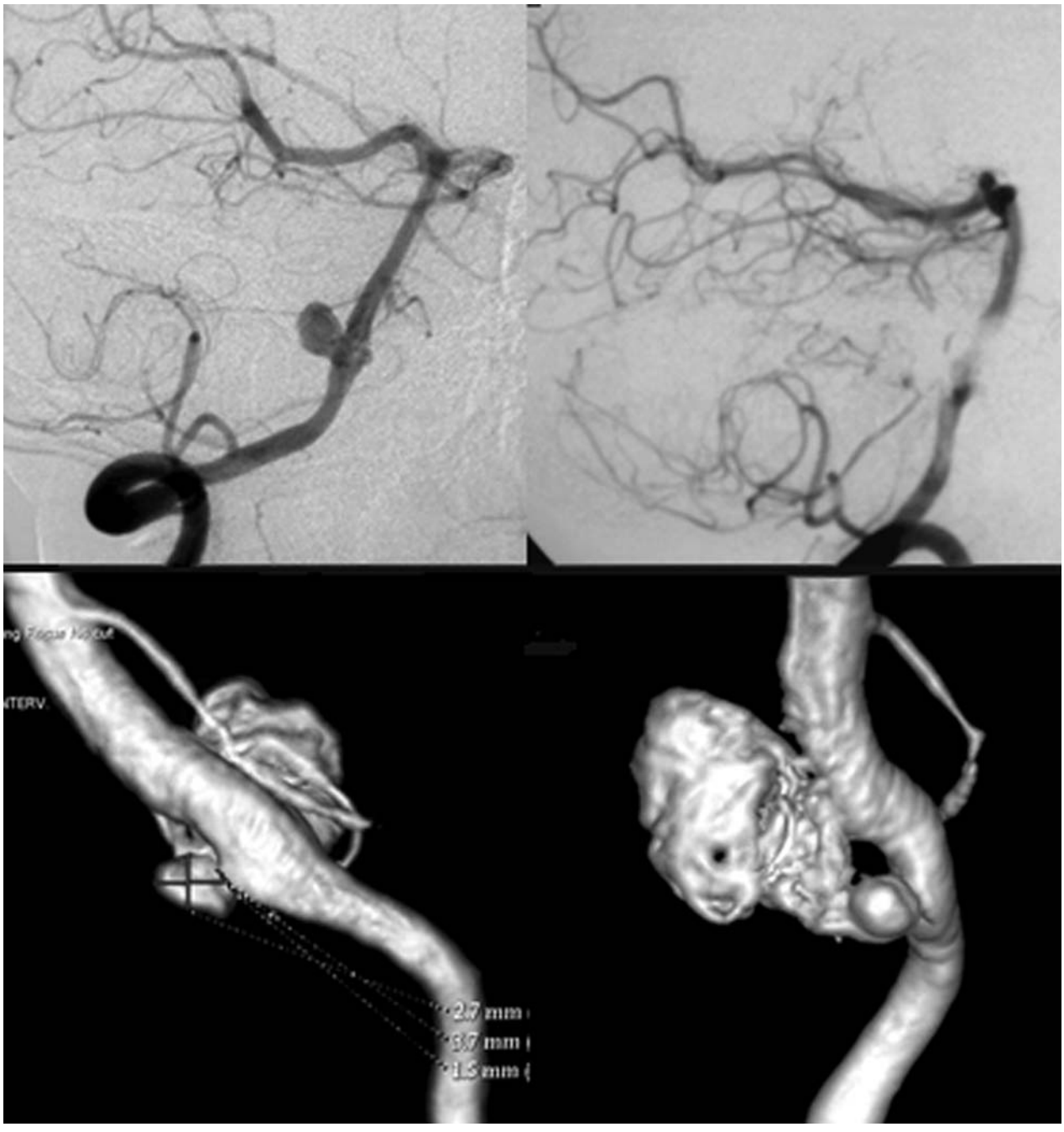

Fig. 1. *Case 1*. The upper row demonstrates the lateral view after vertebral artery injection before and after coil embolisation of the larger aneurysm, the lower row represents the 3D images demonstrating a smaller kissing aneurysm on the opposite side of the non-fused basilar artery

associated with ischaemia of the left cerebellar hemisphere. This occurred presumably owing to vasospastic infarcts (since transcranial Doppler was positive for vasospasm of the posterior circulation) although delayed thrombo-embolic events from the coiled aneurysm cannot be fully excluded as the reason for the delayed symptoms. Three months after embolization the dysarthria recovered and follow-up angiography revealed 20% recanalisation of the basilar artery aneurysm which was treated successfully with repeat coil embolisation without causing any neurological disturbance. The second aneurysm demonstrated a broad neck and could not be treated due to its small size and unfavourable neck condition. The patient was followed up with serial angiography which

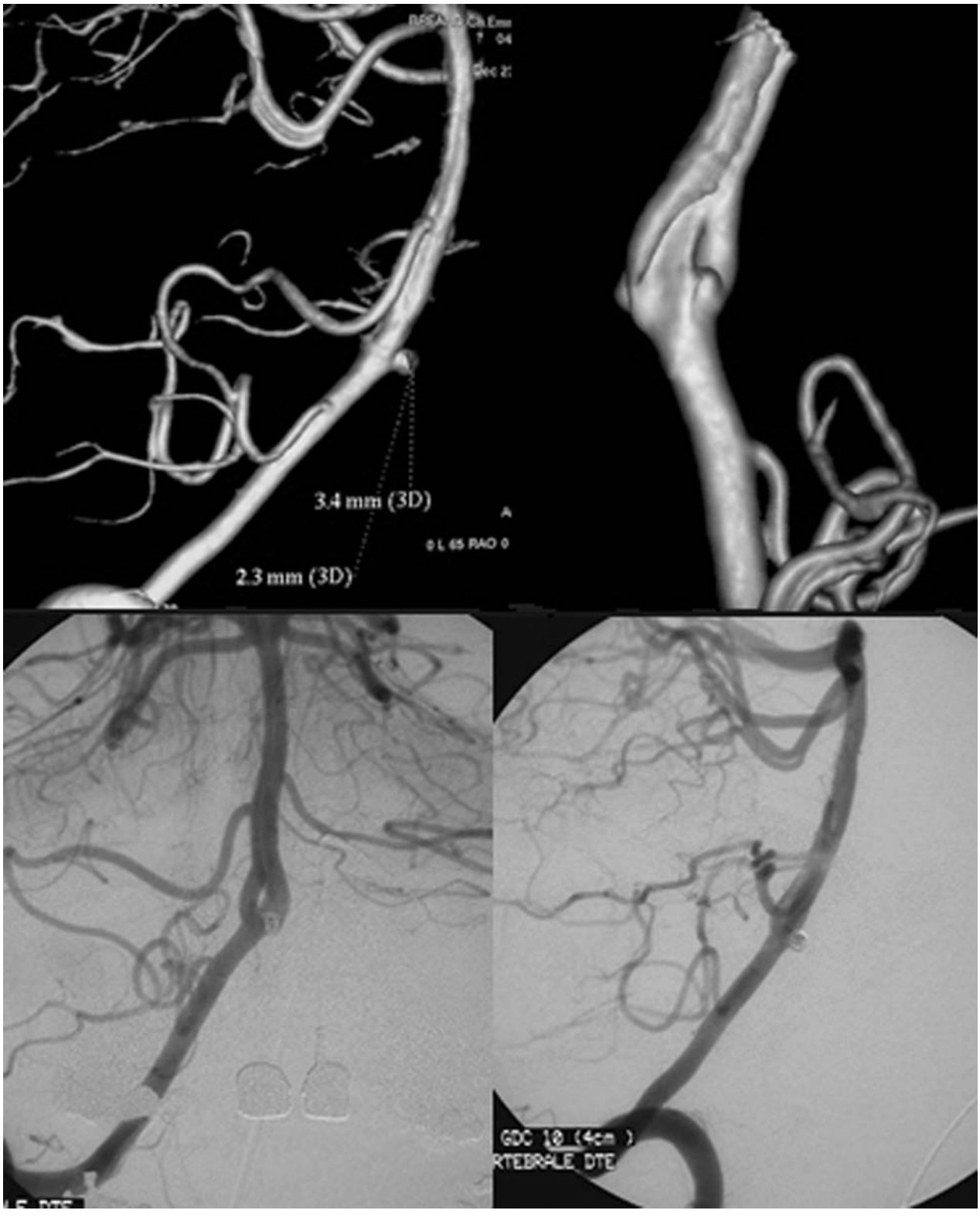

Fig. 2. *Case 2*. In the upper row the pre-interventional 3D images demonstrate an anteriorly directed aneurysm that was subsequently embolised (*lower row*) and a smaller aneurysm ''in statu nascendi'' that was pointing posteriorly, which could not be embolised due to its small size

has demonstrated a stable condition since the last intervention (12 months).

Case 2

A twenty-nine year old man presented with loss of consciousness after a sudden onset of headache and the CT scan demonstrated SAH. Digital angiography revealed a fenestration of the basilar artery associated with two kissing aneurysms at the base of the non-fusion point. One aneurysm projected anteriorly and measured 2.4 mm in diameter while the other, smaller lesion, with a broad neck pointed posteriorly. Another aneurysm was found in the internal carotid artery at the origin of the posterior communicating artery. Embolisation of the larger basilar aneurysm was performed with coils while the patient was in a poor clinical status (Glasgow Coma Scale score 3) and septicemia (Fig. 2). Again, the smaller aneurysm could not be treated due to unfavourable neck conditions and small size. During hospitalisation

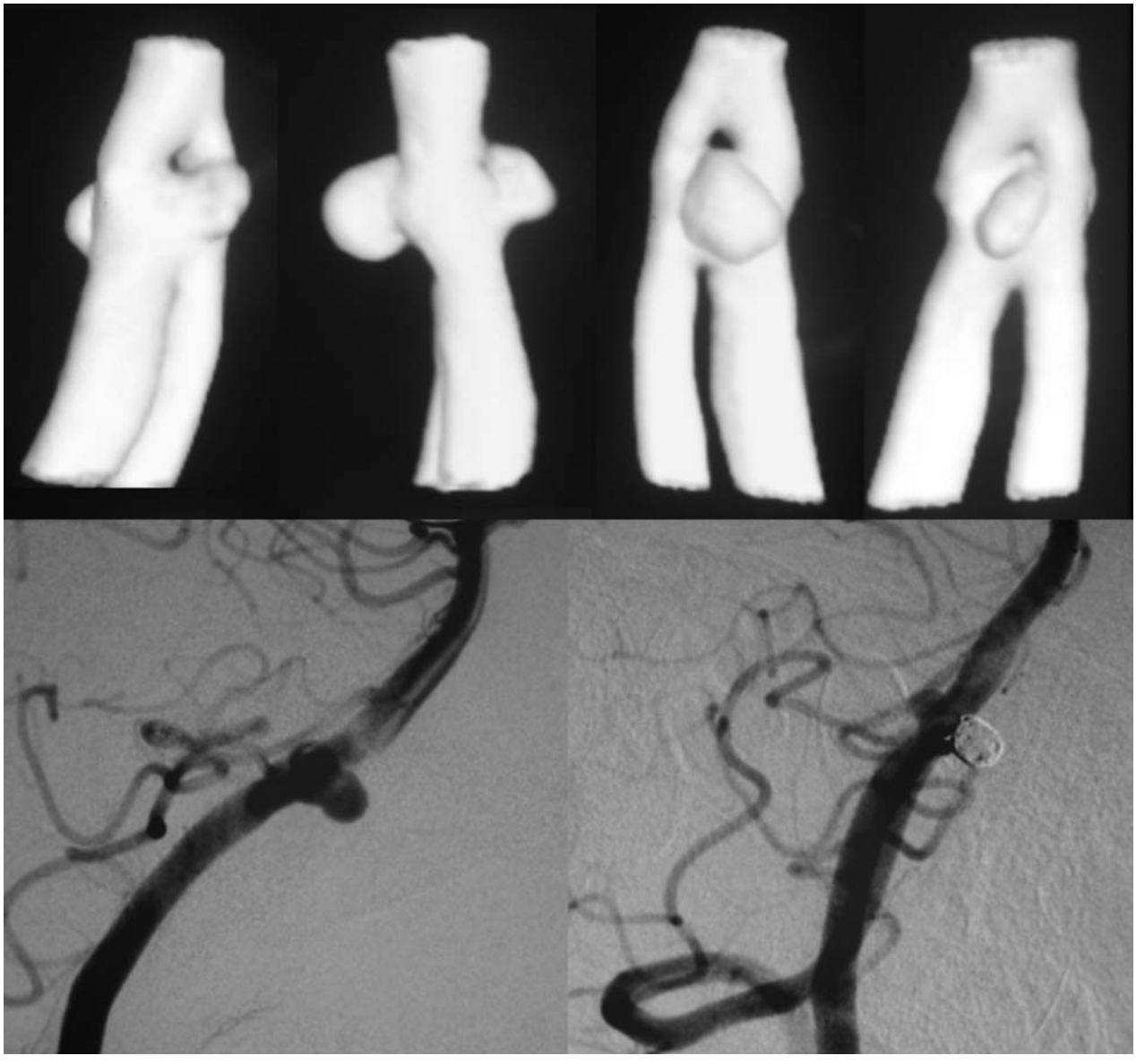

Fig. 3. *Case 3*. The upper row demonstrates the pre-interventional 3D images after injection into both vertebral arteries. Two broad based aneurysms pointing aneriorly and posteriorly are located at the proximal part of the unfused basilar artery segment. Only the larger one could be embolised (*lower row*: pre- and post-interventional images) while the smaller aneurysm had an unfavorable neck to dome ratio which prevented coil embolisation

the clinical condition improved (Glasgow Coma Scale score 15) and he was referred for rehabilitation whilst awaiting follow-up angiography after 12 months.

Case 3

A 34 year-old patient presented with a sudden onset of severe headache without focal neurological deficit and the CT scan revealed SAH. On digital subtraction angiography, a large basilar tip aneurysm with a wide neck as well as a daughter aneurysm (or ''bleb'') and a basilar fenestration with kissing aneurysms pointing anteriorly and posteriorly were demonstrated. The basilar tip aneurysm responsible for the haemorrhage was treated by endovascular means. The patient had an uneventful recovery and subsequently underwent successful embolisation of the larger wide necked kissing aneurysm without complications (Fig. 3). Coiling of the smaller aneurysm, however (dome: 1 mm, neck 2 mm) proved to be impossible, even with a balloon-remodeling technique and stent-assisted coiling was felt to be too dangerous because of the small size of the two parent vessel channels and the communication of the aneurysm with the lower fused part of the basilar artery and the risk of coil protrusion into the unprotected channel. Since there was no dominance of one vertebral artery, the fenestration and the exact location of the aneurysms could be best perceived after bilateral simultaneous injection into both vertebral arteries during 3D angiography.

Discussion

The basilar artery is formed as a result of two simultaneous fusion phenomena both related to regression of the trigeminal arteries. When the embryo is about 4 mm long, the bilateral longitudinal neural arteries are recognisable, bordering the ventral midline non-vascular strip, and are connected laterally at multiple locations with the primitive hindbrain plexus. At this stage, two fusion phenomena take place, one at the ponto-mesencephalic sulcus that is related to the caudal divisions of the internal carotid arteries (ICAs), while the other fusion process involves the longitudinal neural artery system and vertebro-basilar maturation (Fig. 4). Caudal basilar fusion occurs with late trigeminal involution and cranial basilar fusion with early involution, because the trigeminal artery is responsible for the flow changes in the vertebral system during arterial development [2]. The fusion process is typically completed when the embryo is 9 mm long (i.e. around the 5th foetal week); a single basilar artery is formed due to the above mentioned cranio-

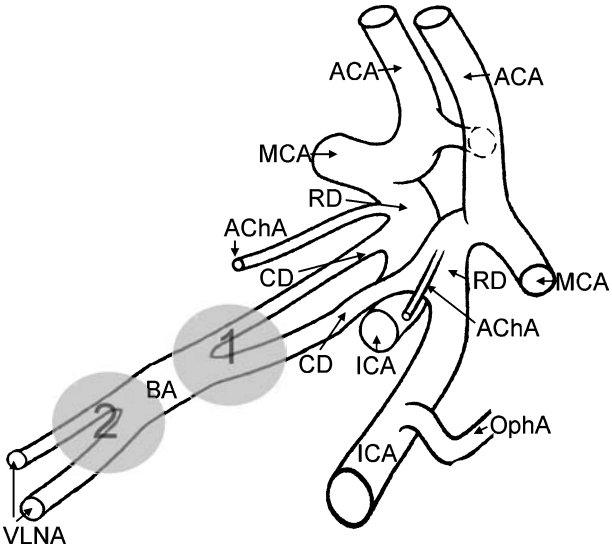

Fig. 4. When the human embryo is about 4 mm long two fusion phenomena take place (*1 and 2*), the first (*1*) at the ponto-mesencephalic sulcus in which the caudal divisions (*CD*) of the internal carotid arteries (*ICA*) fuse to form the tip of the basilar artery (*BA*). The other fusion process (*2*) involves the ventral longitudinal neural arteries (*VLNA*) that form the basilar artery more caudally. *OphA* Ophthalmic artery, *AChA* anterior choroidal artery as the first branch of the rostral division (*RD*) of the ICA, *ACA* anterior cerebral artery, *MCA* middle cerebral artery

caudal fusion of the embryonic longitudinal neural arteries [14, 15]. Failure of fusion of the neural arteries with regression of the bridging vessels between longitudinal arteries is an explanation for the phenomenon of basilar artery fenestration [10]. Given the above mentioned embryology, this term is however incorrectly applied to the condition described in this paper: A *fenestration* is defined as a single artery with two luminal channels. This fenestration can, for example, be due to a nerve or other anatomical structures ''piercing'' the artery and is typically encountered in the vertebral artery or the ICAs in the neck. A lack of fusion of embryologically paired vessels, on the other hand, leads to *segmentally unfused arteries*. This condition can only exist where two embryological arteries fuse during development. Therefore, the basilar artery – as described in this report, or the anterior spinal artery can harbour unfused segments. In addition, *duplications* can occur where the ''double lumen'' is due to two embryologically different vessels that fuse during development; an additional vessel persists, whereas in fenestrations the two lumina correspond to a single artery. Duplications can be encountered in the ICA (segmental agenesis of the first ICA segment with reconstitution of the distal ICA via different arteries from the ascending pharyngeal artery system), the anterior cerebral arteries (ACAs) with duplicated vessels and persistent infraorbit-

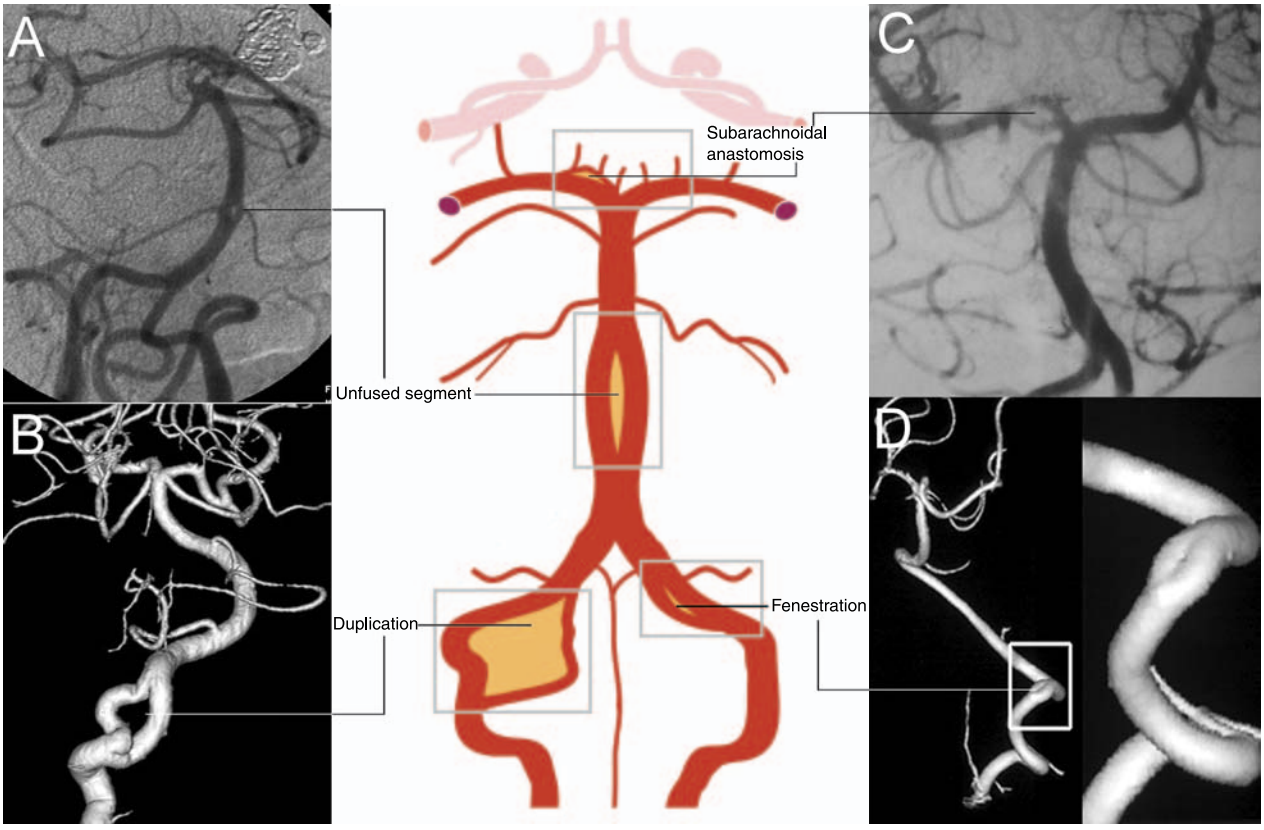

Fig. 5. This drawing demonstrates the different forms of ''double lumen'' that can be encountered: An *unfused segment* (A) is the result of an incomplete fusion of two embryological vessels (in case of the basilar artery: the paired ventral longitudinal neural arteries). A *duplication* (B) refers to persistence of an additional embryological vessel, i.e. two separate vessels are present (in this example the lateral spinal artery and the vertebral artery). Extra-cerebral anastamoses between perforators can also form a double lumen and is established from a rete of perforators present in the embryo (C). A *fenestration* is defined as a single artery being ''pierced'' by various anatomical structures (most often nerves) thereby creating two separate luminal channels of the same artery (D)

al origin of the ACAs, or at the vertebrobasilar junction. Here, the duplication recruits two separate vessels (the lateral spinal artery and the vertebral artery), one of which enters the spinal canal, the other remaining in the vertebral canal. Finally, *extracerebral anastomoses between perforators* can lead to the phenomenon of a double lumen in a single artery. This condition can be encountered both close to the anterior and posterior perforating substance and is established from the embryological rete of perforators (Fig. 5).

Non-fusion of the basilar artery usually involves the lower half of the vessel because of incomplete fusion of the longitudinal arteries in the cranio-caudal direction, although distal non-fusions have been reported and are very rare (one instance in 2161 patients studied by Trah-Dinh *et al.*) [9].

Since the basilar artery develops by the midline fusion of the paired ventral longitudinal arteries, multiple variations of the vessel will occur depending on the fusion process of the longitudinal arteries and contribution of the segmental arteries (axial vascular system). Complete unfused basilar artery, segmental unfused basilar artery (pseudo-fenestration), or segmental basilar agenesis (with or without trigeminal persistence) are anatomical variations (i.e. non pathological) related to the pattern of fusion [2]. Owing to their embryological development, both limbs of the unfused basilar artery carry brainstem perforating arteries to their respective sides. Therefore, vessel sacrifice of one limb of the pseudo-fenestration may not alter the haemodynamics of the distal basilar artery but there is a high risk of unilateral brain stem stroke due to occlusion of perforators.

Although rare, arterial fenestrations and unfused arterial segments can be associated with intracranial saccular aneurysms [16, 17]. Of 59 aneurysms of the vertebrobasilar junction reviewed by Campos *et al.* [8], 35.5% were associated with a non-fusion of the proximal basilar artery. Basilar artery non-fusion can also be associated with other embryological development defects such as pituitary duplication [18].

The lateral walls of the unfused artery have a normal intrinsic architecture; however, the media layer is absent at the base of the medial wall and in addition, the subendothelium is thin and there is discontinuity of the elastin layer [19]. It is interesting that cerebral artery bifurcations have similar structural anatomy [19, 20]. A morphological postmortem study of an unfused basilar artery aneurysm confirmed that the muscular layer of the basilar artery is also absent adjacent to the aneurysm in this location [21]. We believe that the association of arterial variants such as incomplete fusion points to an incomplete maturation process of the arterial wall. The lack of cell selection that such patterns imply may preserve ''weaker'', i.e. less matured endothelial cells, which will later develop arterial aneurysms when secondary, revealing, triggers such as haemodynamic stress are present [2].

Two distinct aneurysms arising from a segmentally unfused basilar artery have been reported in five patients: Saatci *et al.* report on a patient who presented with subarachnoid hemorrhage and in whom two aneurysms arising from the proximal part of the unfused segment were found. Both of them were treated successfully with coils. Tasker and Byrne [13], Yoon *et al.* [12], Islak *et al.* [4] and Campos *et al.* [8] reported one patient each in their series of five, four, 10, and 20 patients respectively. Two of these latter four patients were treated with coils successfully and without complications, one patient belonged to a surgical series and one patient was not treated. Our three patients are taken from a series of 35 patients with basilar artery non-fusions associated with aneurysms (of a total of 197 aneurysms of the posterior fossa). Given the experience with this condition in the literature, and our experience, we conclude that the occurrence of kissing-type aneurysms at a non-fusion site might be as high as 10% and should, therefore, be sought for specifically. We think that three-dimensional angiography is an extremely helpful tool to establish the diagnosis, which can be overlooked when using biplane angiography, even when using multiple angulations.

The surgical treatment of these aneurysms is difficult due to their relationship to the cranium, lower cranial nerves, and the complex surgical approaches to this region [5, 22]. In the surgical series of Campos *et al.* [8], complete occlusion of the aneurysm was achieved in 70% of cases and incomplete protection in 15%. Aside from one death, more then half of these patients had transient lower cranial nerve palsies and one a permanent neurological deficit [8].

Endovascular embolisation is an alternative to surgery for aneurysms related to an unfused basilar artery [4, 13, 23, 24]. Wide necked or unfavourably shaped aneurysms can be treated using balloons in both vertebral arteries permitting the remodeling of the aneurysm neck with coils [25]. In the series of Islak *et al.* [4], the endovascular treatment of 11 patients with unfused basilar artery aneurysms enabled an occlusion rate of 95%. There was one instance of aneurysm re-growth after one year re-treated successfully. However, it has to be taken into account that both limbs of the unfused basilar artery have to be preserved. Aneurysm re-growth might occur due to the unfavourable haemodynamics at the site of the unfused segment and also because the aneurysms often have a broad neck. We were only able to treat the larger of the two kissing aneurysms in each patient because of unfavourable dome to neck conditions and therefore obliged to closely follow up the patients to ensure timely treatment if there is a change of morphology.

In our series of three patients some particular points have to be highlighted: a) In all patients, an additional aneurysm was present (located on the MCA, the posterior communicating artery and the basilar tip respectively, b) the communication between the aneurysms at the non-fusion site and the parent artery always involved both limbs and was, therefore, extremely broadly based with an unfavourable neck-to-dome ratio, c) visualisation of both aneurysms and the unfused segment was best achieved with a bilateral simultaneous injection via both vertebral arteries so as to avoid inflow effects of the contralateral vertebral artery when both vertebral arteries were of equal diameter, and, d) in all cases the second aneurysm was too small to be occluded with coils and in fact resembled more an aneurysm ''in statu nascendi''.

We conclude that ''kissing'' aneurysms located on unfused basilar artery segments may be more common than previously thought, that 3D rotational angiography (ideally with bilateral injections into both vertebral arteries) is the diagnostic modality of choice in treatment planning, and that advanced endovascular techniques including balloon remodeling might be necessary to treat these aneurysms.

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Comment

Even if this observation is ''more common than previously thought'' it will remain for the readers ''exceedingly rare''. The clarity of the text and the quality of the pictures make this paper a worthy addition to the literature.

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