Saccular aneurysm associated with segmental duplication of the basilar artery

A morphological study

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Saccular aneurysm associated with segmental duplication (also called “fenestration”) of the basilar artery is an anomaly that results from an embryonic fault. Reports of the treatment of the aneurysmal component have only recently appeared in the neurosurgical literature, and little has been written on the morphology of this anomaly. This study answers the need for information about its structure to the extent permitted by the examination of a single specimen.

The specimen was obtained at postmortem examination. A cast of its interior features was made with a synthetic rubber. After the cast was removed, the entire anomaly was serially sectioned for histological study. Defects in the wall of the basilar artery were seen microscopically at each end of the fenestration. At the extensive proximal defect, a saccular aneurysm arose that bulged into the window between the two limbs of the segmental duplication and also presented dorsally and ventrally. It had fatally ruptured. The manner in which the fenestration was formed by intraluminal septa was also revealed.

KEY WORDS • basilar artery • segmental duplication • saccular aneurysm • morphological study

The treatment of saccular aneurysm associated with segmental duplication (also called “fenestration”) of the basilar artery is a recent advance. Reports of the arteriographic appearance of this anomaly and the management of its aneurysmal component date only from 1976.1,3-7,9,10 Basilar artery fenestration found at autopsy, without an attendant aneurysm, has a longer history,2,11 but the only morphological study of the condition appears to be that of Crompton.2 He said: “Histological examination of these fenestrations has revealed a defect in the media at the proximal end.” Aneurysm formation at that site was not mentioned, but one was shown in an accompanying drawing of the cerebral arteries. Morphological study of an entire basilar artery fenestration, with aneurysm formation, has remained to be done. That is the subject of this paper.

Clinical Material

The patient was a 24-year-old white man who was found dead in bed. There was no obvious cause of death. In his youth the spleen had been removed following trauma. Several days before death he was said to have had a “sinus headache.”

Postmortem examination was without abnormality save for abdominal scars and a swollen brain, where intraventricular and subarachnoid hemorrhage was found. The source of the bleeding was described as a “ruptured aneurysm” located on the dorsal aspect of the basilar artery and presenting to the right at the pontomedullary junction. Otherwise, the cranial arteries were normal.

Embryological Substrate

Padget8 has shown that the basilar artery is formed by fusion of the paired, laterally placed, longitudinal neural arteries. Fusion usually occurs during growth of the embryo from a crown-rump length of about 5 to 9 mm. During that time the basilar artery is described as being “irregular and marked by many islands.”9 The islands are regions where fusion of the longitudinal neural arteries has yet to occur. An occasional island is present even at 12 mm, and in some cases fusion is not completed. Where the longitudinal neural arteries remain separated, segmental duplication results. It is found in about 5% of some autopsy series11 and can occur anywhere along the course of the artery.
External Appearance

On the dorsal aspect of the basilar artery, just cephalad to the vertebrobasilar junction, there was a saccular outpouching 5 mm in diameter (Fig. 1). It had ruptured. An opening, tricorn in shape and 2 mm wide, was seen at the top of the fundus. Lying in the surrounding blood clot was a cap-like structure of the same shape as the sac and attached to it by a thin strand of tissue. The cap was gray, thin, and blood-stained on its inner surface. Sections of the cap showed it to be composed of delicate reactive connective tissue containing hemosiderin, which indicated that the aneurysm had bled previously. A midline groove in the artery, covered by adventitia, extended 3 mm cephalad from the outpouching. This segment of the vessel was broader than it was distally.

The ventral surface of the basilar artery also displayed a saccular protrusion. It was smaller than the dorsal sac. A midline groove was also present ventrally, and it too extended cephalad 3 mm from the protrusion. Beyond that the artery was normal.

Cast of the Specimen’s Interior

A cast (Fig. 2) of the interior of the specimen was made by injecting a synthetic rubber (Permlastic)* into the lumen of the basilar artery under a pressure of 120 mm Hg. Owing to the rupture, the Permlastic oozed from the dorsal sac and vitiated its true portrayal. After solidifying, the cast was removed. Some unexpected details were revealed. There was actually only one aneurysm. It was located at the proximal end of the fenestration and had an oval configuration (Figs. 2 and 3). The rounded and apparently separate dorsal and ventral sacs noted externally were revealed to be parts of a larger aneurysm. The two sacs joined cephalad to form that section of the aneurysm that bulged into the window between the two limbs of the segmental duplication. This finding suggested that the aneurysm originated from a curvilinear orifice where the basilar artery bifurcated. In short, this was a carinal aneurysm.

The cast also demonstrated the fenestration as it might have appeared in life. The space between the limbs of the segmental duplication was 3.7 mm in length, widened to 1.5 mm, and was asymmetrical owing to the right branch being larger. The grooves seen externally on the limp specimen signalled the fenestration. An x-ray film of the cast resembled published arteriograms of the anomaly.5,10

Histological Examination

Serial sections were obtained every 25 μ proceeding from the vertebral arteries to the normal basilar artery distal to the anomaly. Every fifth section was stained with hematoxylin and eosin or elastic trichrome. The

* Permlastic (light-bodied) is manufactured by the Kerr Division of Sybron Corp., Romulus, Michigan. It is used mostly for making dental impressions. The manufacturer states that this formula has a dimensional stability of 0.4% at 24 hours. A change of that degree was not detectable in a cast this small, which was measured to 0.1 mm. Permlastic separates from moistened arterial wall with minimal intimal damage. Lead peroxide used in its manufacture makes it radiopaque.
Aneurysm in duplicated basilar artery

FIG. 3. Drawing of the segmental duplication and associated aneurysm as reconstructed from study of the cast and histological sections. This cut-away projection does not define the dorsal opening in the artery or show the adventitia spanning the fenestration.

vertebral arteries united normally to form a short confluent section of basilar artery. Throughout its course, except for the aberrations at each end of the fenestration, the basilar artery displayed normal intima, internal elastic lamina, muscularis, and adventitia. At a point 1.8 mm from the vertebrobasilar junction, as measured on the cast, an opening developed on the dorsal aspect of the artery. It was midline and widened to at least 1.6 mm. Surmounting that opening was the dorsal component of the aneurysm. Its histological features were not unusual for an aneurysm. Fibrointimal pads were present at the orifice of the sac; reduplicated internal elastic lamina and irregular segments of smooth muscle were also noted at the orifice and extended for a short distance into the neck of the sac. Hyalinized connective tissue, which thinned toward the rupture in the dome, composed the wall of the sac. The orifice did not close cephalad, but rather continued into the aneurysmal wall extending in that direction.

The ventral opening was shaped like the dorsal opening and began 2 mm beyond the vertebrobasilar junction. It was about 1 mm wide and, like the dorsal opening, did not close distally where it was continuous with the cephalad projection of the aneurysmal wall. The histology of the ventral sac was similar to the dorsal outpouching. A defect in the media only was present for a longitudinal distance of 200 μm proximal to the ventral opening (Fig. 4). The intima and internal elastic lamina gradually thickened over a slightly longer distance, 375 μm. Laterally, the wall of the basilar artery was histologically normal.

Septa within the lumen of the basilar artery were also noted. These were vestiges of the paired embryonic longitudinal neural arteries, and marked the beginning of the segmental duplication. The septa appeared first about 1 mm cephalad to the start of the dorsal opening and were on the right side of the orifices (Figs. 3 and 5). The septa, continuous with the arterial wall, were composed of smooth muscle, thickened internal elastic lamina, and intima covered by endothelium. They entered both dorsally and ventrally. Their progress inward

was not equal from each direction nor did they start in the same longitudinal plane. Within about 1 mm they united to form the medial wall of the right branch of the duplication, which then proceeded cephalad as a separate vessel. Formation of the left branch of the duplication by entering septa was similar to that on the right but began more cephalad. These septa developed to the left of the orifices and within 1 mm formed the left branch. Before that was completed, the aneurysm was washed by blood flowing mainly on the left side of the basilar artery. This may account for the tilt of the dorsal sac to the right seen at autopsy. The septa on the

FIG. 4. Photomicrograph of the ventral aspect of the basilar artery at the beginning of the orifice. A myointimal pad is present on the left of the rift, and thickening of the internal elastic lamina is seen on the right. Arrows define the length of the muscular defect. Elastic trichrome stain, × 25.5.

FIG. 5. Drawing of the lumen of the basilar artery at the proximal end of the fenestration as perceived from examining serial sections through that region. The septum on the right forms before that on the left.
branches of the segmental duplication reunite. A medial defect
the cast. Elastic trichrome stain, x 23.

The cut through the wall dorsally was made in order to remove
the defect is a myointimal pad and internal elastic lamina.

is present on the ventral aspect of the vessel
branches reunited, however, revealed thinning of the
common adventitial coat. Study of the area where the
midline aneurysm and the segmental duplication. There the embryonic vessels per-
from the longitudinal neural arteries save for the seg-
multiple at the proximal end of the fenestration and pro-
sistent as paired channels. Histologically, all arteries were
were histologically identical to those on the right.
left were histologically identical to those on the right.
proximal end of the fenestration, then, there were three compartments: the right and left branches
Histological examination of the two separated
branches showed normal vascular morphology and a
common adventitial coat. Study of the area where the
branches reunited, however, revealed thinning of the
muscular coat and actual absence of it over a longitudi-
distance of about 50 μ. The intima overlying this area was slightly thickened, but the internal elastic
lamina was normal (Fig. 6). The thinning, ventrally
located and slightly off center, occurred where the mus-
cle layers of the two branches met. It is the other, the
distal, defect the embryology foretold. This defect re-
sembles that proximal to the aneurysmal orifice on the
ventral aspect of the basilar artery (Fig. 4). There were
no intraluminal septa distally.

In summary, the basilar artery was formed normally
from the longitudinal neural arteries save for the seg-
mental duplication. There the embryonic vessels per-
persisted as paired channels. Histologically, all arteries were
normal except at each end of the fenestration, in both of
which locations there was a defect in the vessel wall.
An oval-shaped aneurysm had formed at the extensive
proximal end. The explanation is seen to involve
many interlacing factors: location and size of the mural
defect; configuration of intraluminal septa, if present;
hemodynamic forces; and structural changes in the
vessel owing to age. In that last regard, this patient is
the youngest so far recorded.

Embodyd in the previous reports, and enhanced by
our findings, is notice that the radiographic definition
of this anomaly could be difficult and treatment of
the aneurysm formidable. Becker and Hamilton1 and
Peerless8 have warned of the possible presence of an
aneurysm in any case of fenestration. Our findings
support that. The saccular aneurysm can arise exten-
sively at the proximal end of the fenestration and pro-
ject in several directions. A defect in the muscular coat
at the cephalad end of the fenestration indicates that
an aneurysm could, under suitable conditions, form
there also.

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