

Dysplastic Vertebral Artery With Paradoxical Infarction

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SUMMARY: *A rare abnormality of the vertebral artery is described in a 63-year-old male. The right vertebral artery consisted of several minute endothelial lined channels which failed to perfuse at autopsy. The anterior spinal artery showed areas of muscular media dysplasia. These anomalies were associated with large posterior communicating arteries. The terminal event was precipitated by thrombotic occlusion of the left vertebral artery. The embryological aspect of these anomalies are discussed.*

RÉSUMÉ: *Nous décrivons une rare anomalie de l'artère vertébrale chez un mâle de 63 ans. L'artère vertébrale droite comprenait plusieurs fins canaux couverts d'endothélium qui ne purent être perfusés à l'autopsie. L'artère spinale antérieure montre quelques zones de dysplasie de la média musculaire. Ces anomalies étaient associées à de grosses artères communicantes postérieures. L'évènement final fut précipité par une occlusion thrombotique de l'artère vertébrale gauche. L'aspect embryologique de ces anomalies est discuté.*

INTRODUCTION

In view of the complex development of the vertebral-basilar arterial system, it is not surprising that developmental anomalies occur in these vessels. Considerable differences in the caliber of the vertebral arteries have repeatedly been described (Sieglbauer, 1958; Priesching, 1956; Hutchinson and Yates, 1956; Schmitt, 1973). This asymmetry in size is usually maintained throughout the course of the vertebral arteries (Hutchinson and Yates, 1956). True hypoplasia or aplasia of vertebral arteries is not uncommon (Singh and Singh, 1971; Tsai, et al., 1975). Krayenbühl and Yasargil (1957), described an incidence of 0.75% in 400 angiographically evaluated cases. These two abnormalities are frequently associated with persistent embryological vessels such as the trigeminal or hypoglossal artery (Szdzy and Lehmann, 1970; Perryman, et al., 1963). Less commonly reported developmental anomalies consist of fenestration (Takahashi, et al., 1970; Kowada, et al., 1975), partial duplication of the vertebral artery (Kowada, et al., 1973), and non-union (Schmitt, 1973; Berry and Anderson, 1910). A revival of interest in these anomalies has taken place recently because they can now be demonstrated angiographically during life and may cause clinical symptoms, particularly if acquired pathology is superimposed.

The present case demonstrated a minute right vertebral artery that failed to perfuse at autopsy. Histological examination revealed a developmental anomaly which is best described as dysplasia. This anomaly was associated with thrombotic occlusion of the other vertebral artery. Clinically the patient presented with a right posterior fossa mass and acute hydrocephalus.

CASE REPORT

A 63 year old gold-miner was well until January 15, 1979, when he developed dizziness, nausea and vomiting. He was admitted to a local hospital on January 17, with a diagnosis of gastroenteritis and severe dehydration. No neurological abnormalities were noted. Over the next 48 hours he became comatose with elevated blood pressure, irregular breathing and generalized hyper-reflexia. Bilateral temporal burr holes were performed. There was no subdural blood clot but the brain appeared swollen. He was given mannitol and dexamethasone and transferred to Toronto General Hospital on January 19. On arrival, he was comatose with apneic spells. Pupils were in mid-position reacting sluggishly to light. Corneal and oculo-cephalic reflexes were present. The limbs were spastic with bilateral Babinski signs. Skull x-ray was not informative. The CT-scan demonstrated dilated ventricles and an avascular mass in the right cerebellar hemisphere with compression and displacement of the fourth ventricle to the left. A ventricular drain was inserted. Post-operatively he improved slightly. He showed spontaneous limb movements and triggered the ventilator. On January 21, the patient deteriorated. All brainstem reflexes were absent. On January 25, his limbs were noted to be flaccid. He developed pneumonia and died on January 31, 16 days after onset of symptoms.

AUTOPSY FINDINGS

Gross Examination. The immediate cause of death was bilateral bronchopneumonia.

The relevant findings were confined to the head and neck. The cerebrum appeared normal, but the cerebellum and the pons were swollen. There were hemorrhagic infarcts involving the posterior inferior right cerebellar hemisphere and the mid-portion of the same area of the left hemisphere (Fig. 1). The left vertebral artery contained a large atheromatous plaque and an occluding thrombus proximal to the exit of the posterior inferior cerebellar artery (PICA) (Fig. 1). The right vertebral

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artery was minute (Fig. 1) and retrograde infusion via the basilar artery demonstrated complete occlusion proximal to the origin of the right PICA. Both PICAs were patent and perfused readily. Both posterior communicating arteries (PCAs) were unusually large. There was no persistent hypoglossal or trigeminal artery. The internal carotid arteries showed minimal degree of atherosclerosis.

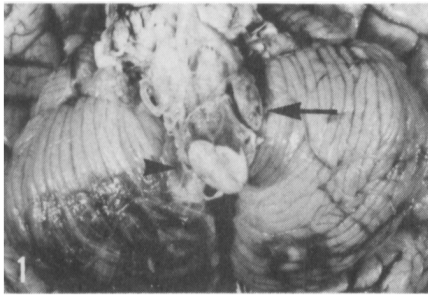


Figure 1 — Hemorrhagic infarction of cerebellar hemispheres in the distribution of the posterior inferior cerebellar arteries. The left vertebral artery shows atheroma with thrombotic occlusion (arrow). The right vertebral artery is minute (arrowhead).

Coronal sections of the fixed cerebral hemispheres showed only the needle-tract leading into the anterior horn of the right lateral ventricle. The ventricles were not dilated. Sections of the right and left cerebellar hemispheres confirmed the presence of hemorrhagic infarctions in the distribution of the PICAs. In addition, there were several small cortical and white matter infarcts in the distribution of both superior cerebellar arteries (SCAs). Sections of the brain stem revealed no gross lesions.

MICROSCOPIC EXAMINATION

Examination of the cerebellar lesions revealed the presence of hemorrhagic infarcts containing lipid and hemosiderin laden macrophages and marginal astrocytic reaction consistent with two weeks' duration. Microscopic infarcts approximately one week old were found in the distribution of the posterior cerebral arteries and the posterior central perforator branches. Likewise, the brain stem showed microscopic infarcts of the right central tegmental tract, the left basis pontis, the right inferior cerebellar peduncle and the right corticospinal tract of the medulla, all approximately one week old. More recent minute infarcts were found bilaterally in the distribution of the anterior inferior and the superior cerebellar arteries.

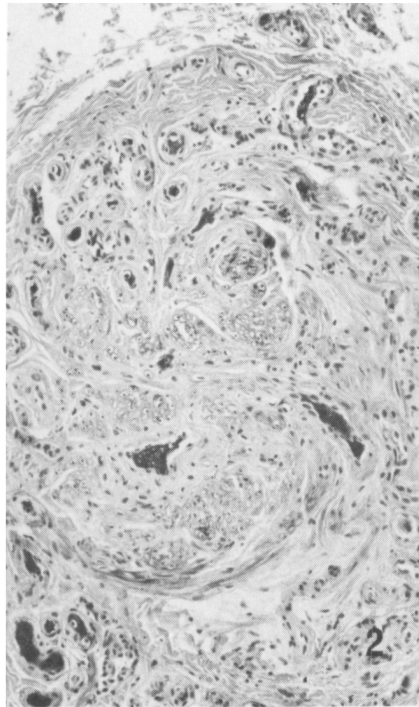


Figure 2 — The right vertebral artery at the level of C5 showing multiple small capillary-sized channels, each with a surrounding cuff of smooth muscle. Haematoxylin-Eosin, X144.

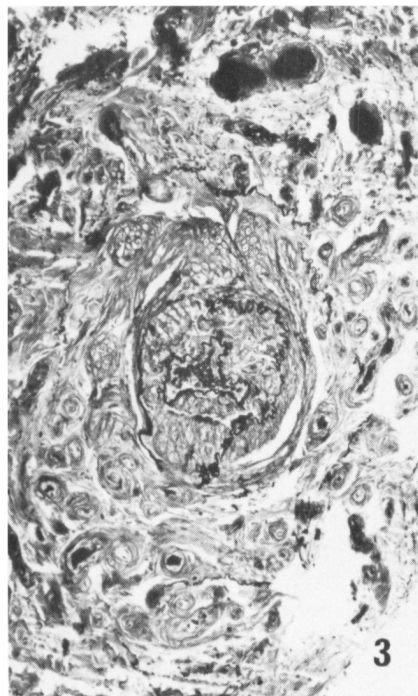


Figure 3 — The right vertebral artery at the level of C2. In the center there is incomplete formation of a muscular artery. Van Gieson Elastica, X144.

The intracranial portion of the left vertebral artery demonstrated an organized thrombus, approximately two weeks old. Its cervical portion was structurally normal and showed no atheroma. The right vertebral artery, below the entrance into the transverse foramen of C₆, consisted of a minute muscular artery of normal structure with no evidence of acquired pathology. Multiple sections taken between C₆ and C₂ showed an unusual malformation. The artery had a well defined outline, but a continuous lumen could not be demonstrated. Instead, the cord consisted of convoluted endothelial-lined channels, each surrounded by a cuff of normal smooth muscle cells (Fig. 2). However, within this segment, at the C₄ and C₂ levels, there was an attempt at forming a central lumen (Fig. 3). Above the C₂ level, the vessel progressed to become a minute normal muscular artery.

The anterior spinal artery, at C₂ and C₆ levels, had a minute lumen, surrounded by a thick dysplastic muscular lamina (Fig. 4).

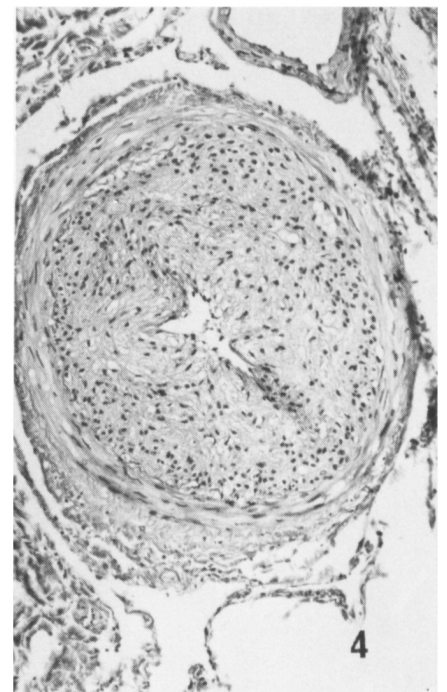


Figure 4 — Spinal artery at the C6 level, showing dysplasia of muscular media. Haematoxylin-Eosin, X144.

DISCUSSION

The present case demonstrated several developmental anomalies of the intracranial vasculature, such as dysplasia of the right vertebral and anterior spinal arteries and unusually

large posterior communicating arteries. The latter finding is indicative of persistence of the embryonic state in which the posterior cerebral arteries arise from the internal carotids via these large caliber posterior communicating arteries (Padget, 1948). It can be assumed that the vertebral-basilar system was at least in part supplied by the carotids prior to the thrombotic occlusion of the left vertebral artery after which it was solely dependent on the supply from the carotids. The distribution of the infarcts supports this assumption, as they involved areas which were located at the distal limits of the anterior circulation. The distribution and ages of the infarcts correspond well with the clinical picture and will not be discussed further.

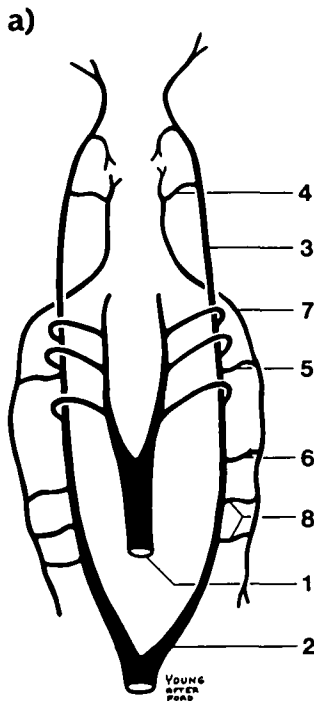
The correlation of the vertebral and anterior spinal arteries anomalies with the different stages of the development of the vertebral-basilar vascular system will provide a basis for their interpretation.

Schmeidel (1932), and later Padget (1948), have provided excellent descriptions of the development of the human vertebral-basilar vasculature. In the 4 mm embryo, the intracranial vertebral arterial system is represented by a plexus of vascular channels which later form the bilateral longitudinal neural arteries (Padget, 1948). (Fig. 5A). These vessels receive their blood supply from the dorsal aortae, i.e., the future carotids, by a number of connecting branches of which the trigeminal artery is the most important at the rostral end. The caudal connecting branches are known as intersegmental arteries (Fig. 5A). These connections normally disappear during later embryonic development, but may persist as anomalous vessels, particularly in association with hypoplasia or aplasia of the carotids or vertebral vessels (Szdzy and Lehmann, 1970; Perryman, et al., 1963; Morris and Moffat, 1956). Thus, in the cervical region of the 4 mm embryo, seven paired segmental arteries arise

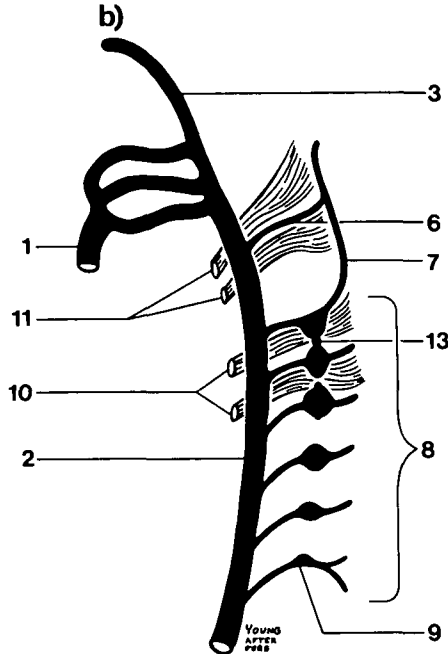
from the dorsal aortae (Schmeidel, 1932). The first pair is known as the proatlantal arteries and the subsequent six as intersegmental arteries, each one associated with the nerve root caudal to it (Fig. 5B). The vertebral artery is formed by a longitudinal anastomosis between these segmental arteries. This anastomosis develops in a definite craniocaudal sequence (Fig. 5B), and the attachments of the corresponding intersegmental arteries to the dorsal aortae are lost. Only the 6th intersegmental artery remains patent as the subclavian artery and the stem of the vertebral artery.

The upper one or two segments of the cervical spinal cord are supplied by the anterior spinal artery, which originates from the medial aspects of the terminal portions of the vertebral arteries (Fig. 5C). Subsequent segments of the spinal cord are supplied by small segmental branches from the cervical segmental arteries, now arising from the vertebral arteries.

4mm Embryo



10mm Embryo



17.5mm Embryo

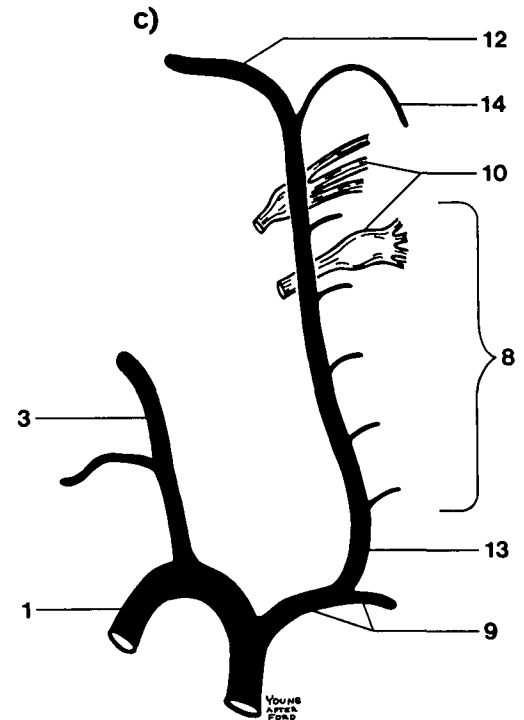


Figure 5 — (A) Diagram of cranial vessels in the 4 mm embryo. (B) Vertebral-basilar system in the 10 mm embryo. (C) Vertebral basilar system in the 17.5 mm embryo. Redrawn from Diagrams of Embryonic Cranial Vessels on pages 9, 15 and 19 in *The Brain Vascular System* by Kaplan, H.A., and Ford, D.H. (1966).

- 1. Ventral aorta 2. Dorsal aorta 3. Internal carotid 4. Primitive trigeminal artery 5. Primitive otic artery 6. Primitive hypoglossal artery 7. Longitudinal neural artery 8. Cervical intersegmental arteries 9. 6th Intersegmental artery (later subclavian artery) 10. Cervical nerve roots 11. Hypoglossal nerve 12. Basilar artery 13. Vertebral artery 14. Anterior spinal artery.

Based on the normal embryonic development of the vertebral artery, it appears that in the present case the craniocaudal fusion between the cervical intersegmental arteries has not been completed normally. A primitive capillary plexus has formed, but the subsequent development of a single endothelial-lined channel has failed. The cervical segments of the anterior spinal artery showed dysplasia of the muscular media with a single small lumen. This presumably represents an arrest of development at a later stage as compared with the anomaly of the right vertebral artery. It is, however, in keeping with the embryological derivation of the anterior spinal artery from the cervical intersegmental arteries.

Persistence of a trigeminal or hypoglossal artery, which in hypoplasia or aplasia of the vertebral arteries, usually provides an anastomosis between the anterior and posterior circulation (Szdzy and Lehmann, 1970; Perryman, et al., 1963), was not found in the present case. Instead, the persistence of large posterior communicating arteries appear to have provided the necessary anastomoses between the carotid and the vertebral-basilar circulations.

To the best of our knowledge, the present anomalies have not previously been described. A solitary case of vasomuscular and media dysplasia of the vertebral artery reported by Rieben

(1973), appears to be a hamartomatous type of malformation of a normal-sized artery.

More detailed examination of hypoplastic vertebral vessels may prove that this anomaly is not unusual. Perhaps some unexplained vertebral artery occlusions in children may prove to be due to similar anomalies (DeVivo and Farrell, 1972).

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