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SHORT REPORT

Acute massive cerebellar infarction associated with craniocervical junction complex malformation: a case report and literature review

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ABSTRACT

We report a case of acute massive cerebellar infarction associated with craniocervical junction (CVJ) complex malformation in a 21-year-old male. Timely surgical intervention prevented the deterioration of his neurological status.

ARTICLE HISTORY

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KEYWORDS

acute massive cerebellar infarction; craniocervical junction complex malformation; posterior fossa decompressive craniotomy; rotational occlusion vertebral artery

Case report

A 21-year-old male was admitted to our hospital due to recurrent dizziness associated with neck rotation for three years. Cerebral computed tomography (CT) in the same day after admission showed no obvious abnormality (Figure 1(A)). Cervical vertebral CT images and three-dimensional reconstruction (Figures 1(B)–(D)) revealed assimilation of C1 with the occiput, atlantoaxial dislocation, and C2–3 and C4–6 fusion with aplasia of C1 and C5 arch. On the left side, the transverse foramen was absent from the atlas (C1) and narrowed in the axis (C2). On the right side, both transverse foramina appeared of normal size. Ultrasound revealed normal cardiac structure and function. At six am on the second day, the patient was found comatose in a prone sleeping position, with his head rotated to the left side. His pupils were of equal normal size but showed poor reaction to light. Emergency CT revealed an extensive low density area in the right cerebellar hemisphere, compression of the fourth ventricle (Figure 1(E)) and dilation of the lateral ventricles (Figure 1(F)). External ventricular drainage (EVD) was immediately performed, followed by a posterior fossa decompressive craniotomy. A cerebellar tonsillar hernia was found during surgery and treated with tonsillar resection. At the 85th hour after surgery, the patient regained consciousness. On the sixth day after surgery, CT angiography (CTA) showed: the right extracranial vertebral artery (VA) entered the spinal canal via the C2 transverse foramen, with its distal intracranial segment displaying significant stenosis; the left extracranial VA entered the spinal canal via the C3 transverse foramen; and the right posterior inferior cerebellar artery (PICA) emerged from the

extracranial segment of the right VA. All arteries showed normal patency without narrowing or occlusion by atherosomatous plaque or thrombosis (Figure 2(A)–(D)). On the 12th day after surgery, cerebrospinal fluid ceased to drain, and the EVD was removed. On the 30th day after surgery, craniocerebral CT showed significant shrinkage of the low density area in the right cerebellar hemisphere and normalization of the lateral ventricle sizes (Figure 2(E),(F)). Although the patient still presented with cerebellar ataxia, he was able to walk with minimal support six months after the surgery.

Discussion

The craniocervical junction (CVJ) is an anatomically complex and functionally unique region which connects the skull and upper cervical spine and is capable of complex motions. Malformations of bone and connective structures can lead to compression of the cervical spinal cord and brainstem, variation in and damage to nearby blood vessels and a series of other pathological changes.¹ We presented a rare case of CVJ malformation with concomitant VA variation.

The blood supply of the cerebellum is unevenly divided into three arteries: PICA, anterior inferior cerebellar artery (AICA) and superior cerebellar artery (SCA). Besides occlusion of one or more of these arteries, massive cerebellar infarction has also been associated with dissection of the VA, embolism from a patent foramen ovale, rheumatic valvular disease, hematologic disturbances and migraines. Our patient is a 21-year-old male with normal cardiac structure and function with no history of such conditions.

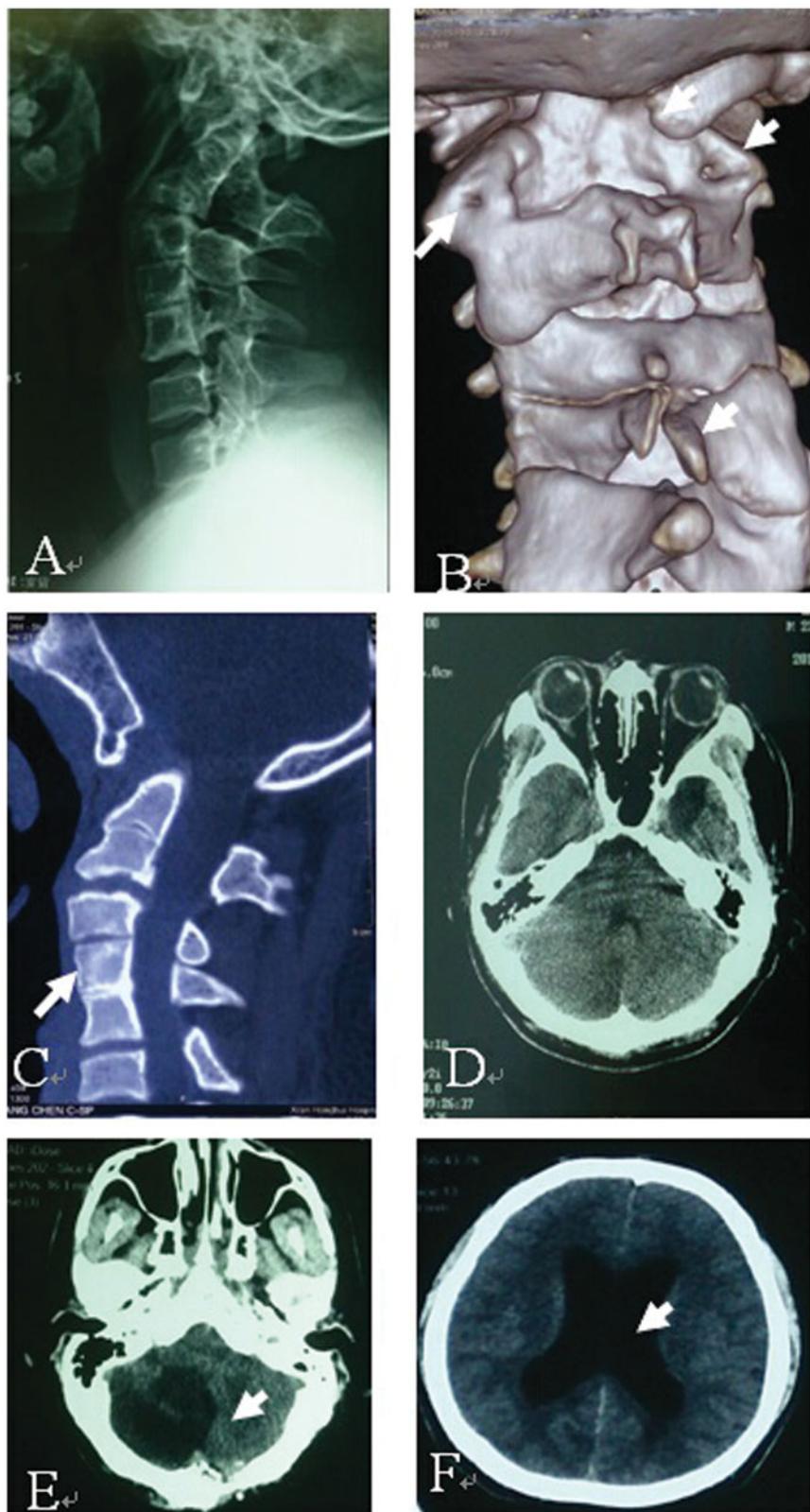


Figure 1. (A) Craniocerebral CT showed no abnormalities. (B–D) Cervical vertebral CT images and three-dimensional reconstruction, showing craniocervical junction complex malformation. Craniocerebral CT showed acute massive cerebellar infarction (E, the white arrow) and dilation of the lateral ventricles (F, the white arrow).

Turning the head to one side may cause narrowing of the contralateral VA at the atlantoaxial level, leading to vascular kinking and stretching, with significantly decreased blood flow or even occlusion (i.e. rotational VA occlusion).^{2,3} Overextension combined with 90° rotation can produce complete VA occlusion above the C2 level. In our patient, the left transverse foramen

was absent from the atlas (C1) and narrowed in the axis (C2), imposing a constant restriction of the left VA that entered the spinal canal at the C3 transverse foramen. This rendered the right VA as the dominant blood supply vessel. Meanwhile, he had significant stenosis at the distal intracranial segment of his right extracranial VA. This made his right VA vulnerable to rotational

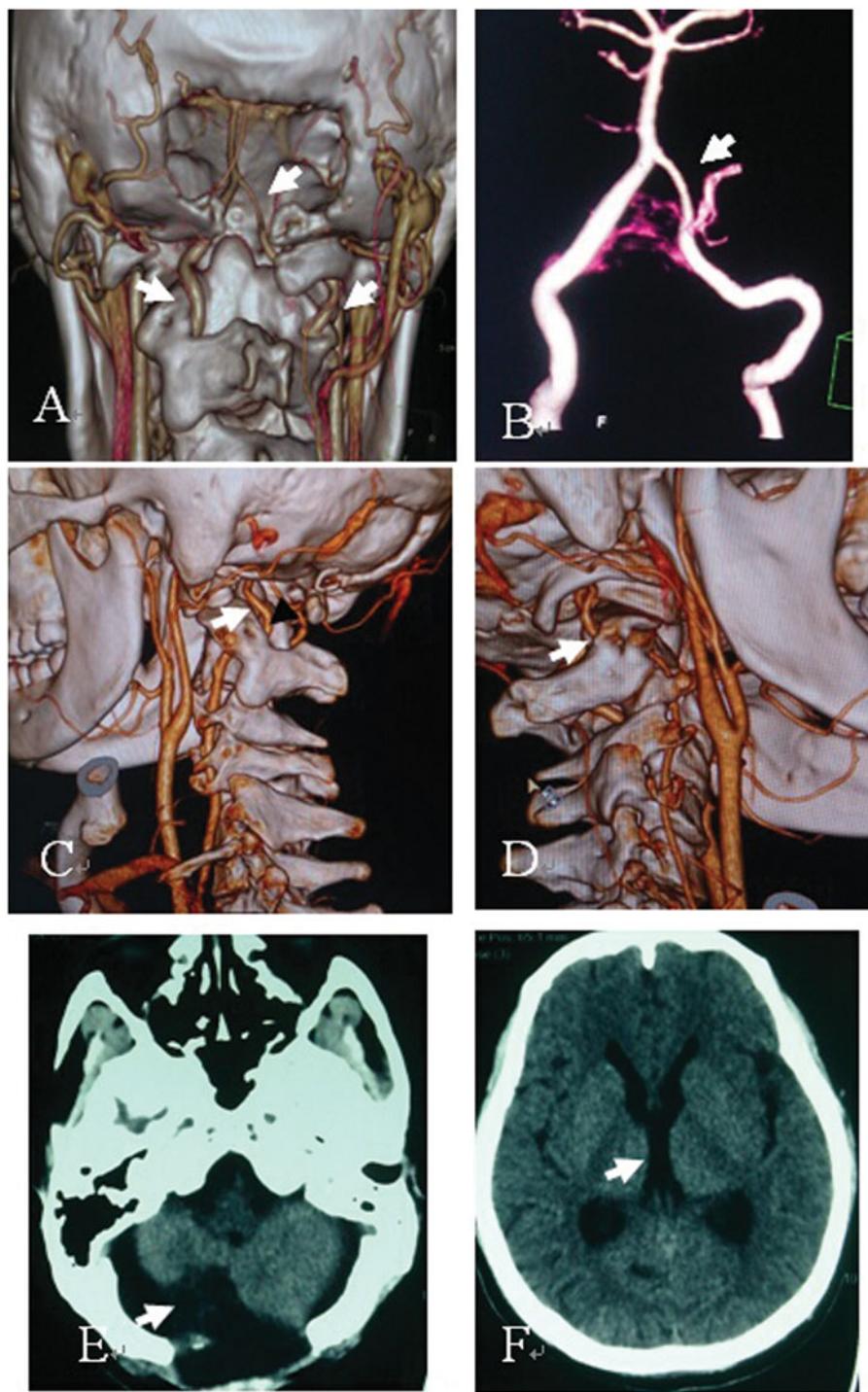


Figure 2. CT angiography showed significant stenosis of the distal intracranial segment of the right extracranial VA (A, the black arrowhead; B, the white arrow) and that the right PICA emerged from the extracranial segment of the right VA (C,D, black arrowheads). Craniocerebral CT showed significant reduction of the low density area in the right cerebellar hemisphere (E) and normalization of lateral ventricle sizes (F).

VA occlusion when his head was rotated to the left, compromising the blood flow in the vertebrobasilar artery territory.

Rapid edema formation following massive cerebellar infarction can lead to life-threatening brain stem compression. Emergent EVD and posterior fossa decompressive craniotomy prevented deterioration of the neurological status in our patient.

Conclusions

CVJ malformations may be concomitant with vertebral artery variation and cause acute massive cerebellar infarction. Timely

surgical intervention can reverse the deterioration of neurological status.

Disclosure statement

The authors declare that they have no competing interests.

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