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Case report

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Giant vertebral artery aneurysm presenting with 'hemiplegia cruciata'



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1. Introduction

Giant aneurysms act by means of their size as space-occupying lesions whereas subarachnoid hemorrhage is rare. Depending on their size and location the huge saccular or fusiform aneurysms of the posterior circulation manifest with varying neurological signs which result from direct compression on the cranial nerves, cerebellum or the brainstem. The syndromes of cruciate paralysis and 'hemiplegia cruciata' are uncommon clinical manifestations of neurological compromise at the cervicomedullary junction [1,2]. Though varied symptomatology of vertebrobasilar aneurysms is known, there is no report till date of posterior circulation aneurysm presenting with hemiplegia cruciata.

2. Case report

A 63-year-old male presented with gradually progressive weakness involving the right hand of 3 months duration. He also developed numbness and tingling paresthesia in both lower limbs which ascended to involve his upper extremities over the next 3 weeks and just prior to admission he noticed stiffness in his left lower limb. On presentation, there was moderate spasticity and weakness predominantly in the right arm and left leg. There was no evidence of muscle atrophy. Pain and thermal sensation disturbances were noted mainly in the right arm, and touch and vibration senses were markedly diminished in the lower extremities. Deep tendon reflexes were exaggerated particularly in the right arm and left leg, and pathological reflexes were also positive on the right arm.

The patient was first evaluated at a local hospital where a magnetic resonance imaging (MRI) brain revealed a $28 \text{ mm} \times 17 \text{ mm}$ lesion in the region of foramen magnum anterior to the cervicomedullary junction. There was significant compression of the brainstem with posterior kinking at the cervicomedullary junction. The lesion was isointense to the brain on T1-weighted sequences and hypointense on T2-weighted images (Fig. 1A–C). CT angiogram brain revealed a giant fusi-saccular aneurysm from the V4 segment of the left vertebral artery in the perimedullary cistern. There was evidence of a large intraluminal thrombus with wall calcification. The right vertebral artery was hypoplastic (Fig. 1D-F). In view of its large size, wide neck and location, the patient was offered stent assisted coiling of the aneurysm which he refused. He was not willing to undertake the risk associated with any surgical procedure. Patient was discharged and came back for review after 6 months. His right upper limb weakness had minimally increased though his lower limb neurological status remained the same.

3. Discussion

Giant posterior fossa aneurysms comprise 5% of all intracranial aneurysms. Clinically symptoms vary, including gait abnormalities, multiple cranial nerve palsies, nystagmus, limb weakness, respiratory dysfunction combined with sleep apnea and hydrocephalus. Ganti et al. [3] reported 8 cases of giant aneurysms of the vertebrobasilar system and divided them into 3 groups based on the location; those at the vertebrobasilar junction with brainstem symptoms, basilar tip aneurysms presenting as mass lesions in the

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Fig. 1. Axial T1-weighted MRI (A) at the level of foramen magnum reveals an isointense lesion situated anterior to the brainstem with an area of hypointensity within it. This lesion appears hypointense on T2-weighted sagittal image (B) and shows enhancement on gadolinium contrast with a flow void signal within it (C). CT angiogram demonstrating a fusi-saccular aneurysm (arrow) from the left V4 segment with intraluminal thrombus and wall calcification (D–F).

floor of the third ventricle and mass lesions in the interpeduncular fossa presenting as a stroke syndrome. The progression of brainstem signs and symptoms has been attributed to the development of intrathrombotic capillary channels that increase the growth of the thrombosed giant aneurysm [4].

The condition named 'cruciate paralysis' by Bell is an infrequent neurological finding [1]. The lesion is situated at the cervicomedullary junction, proximal to the pyramidal decussation. This anatomic and topographic difference sometimes causes unusual clinical manifestations such as cruciate paralysis and hemiplegia cruciata. Characteristically, patients with cruciate palsy present with bilateral paresis of the upper extremities without significant involvement of the lower extremities. When the neural compromise occurs predominantly on one side, spastic palsy on the ipsilateral side of the upper extremity is present, which is associated with spasticity on the contralateral side of the lower extremity, described as hemiplegia cruciata. The neuroanatomical explanation was outlined by Wallenberg [2]. He suggested that a complex somatotopic and anatomical segregation of the corticospinal tracts in the decussation at the cervicomedullary junction resulted in distinct locations for the decussating fibers of the arms and legs. It was presumed that a unilateral injury to the cervicomedullary junction could impinge on the recently crossed corticospinal arm fibers and the uncrossed leg fibers. Even when suspected clinically, cruciate paralysis may be difficult to differentiate from findings associated with the cervical central-cord syndrome. An alternative mechanism to explain this unusual neurological sign is that the corticospinal tract in humans is more important for hand and arm function than it is for lower extremity function. Hence, any injury to the corticospinal tract, even if diffuse, would produce a disproportionately greater upper limb dysfunction than leg dysfunction [5]. In the present case also, the gradual enlargement of the aneurysm would have caused progressive compression on the decussating fibers of the corticospinal tracts resulting in differential involvement of the upper and lower limbs. The presence of intraluminal thrombus and wall calcification may be responsible for the sustained mass effect leading to unilateral injury to the cervicomedullary junction. Another neurological finding uncommonly observed in foramen magnum lesions is an "around the clock" or "inverted U" pattern of upper motor neuron distribution weakness that typically involves the ipsilateral upper extremity, spreading to involve the ipsilateral lower extremity, before involving the contralateral lower extremity and then the upper extremity. This neurological sign also described as "Elsberg phenomenon" is possibly related to impaired venous drainage in the water shed zone of the spinal cord and the traction on the corticospinal tracts produced due to its close proximity to the ligamentum denticulatum.

Direct surgical attack on giant aneurysms of the vertebral artery involves proximal ligation, neck clipping, or aneurysmectomy. Surgical clipping of giant posterior circulation aneurysms is difficult owing to their size, proximity to critical neurovascular structures and presence of an unfavorable neck for clipping. We advocate exploration in most cases as the true nature of the neck is not always appreciated on angiography. This often leads to dramatic improvement of neurological deficits immediately after evacuation of a thrombosed giant aneurysm, quite different from the gradual improvement that occurs after removal of benign tumors in the same location.

4. Conclusion

The clinical history of our patient is a clear example of the diagnostic pitfalls encountered in cases of foramen magnum tumors. In light of the present experience it is suggested that a giant vertebral artery aneurysm should be considered when a mass is observed in the region of foramen magnum accompanied by atypical clinical features.

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