Bilateral dissecting aneurysms of the internal carotid arteries misdiagnosed as skull base tumors: A case report

BIN QI, ZHI-CHENG LU, WEI WU and YI-PING LI

Department of Neurosurgery, First Hospital of Jilin University, Changchun, Jilin 130021, P.R. China

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Abstract. A 45-year-old female presented with a five-year history of intermittent headaches and a two-month history of left hypoglossal nerve palsy. Computed tomography and magnetic resonance imaging of the head revealed space-occupying lesions in the base of the skull with accompanying bone erosion, which were suggestive of skull base chordomas. However, an endoscopic endonasal transsphenoidal biopsy was also performed and pathological analysis of the lesion suggested a thrombosis. Cranial magnetic resonance angiography revealed old dissecting aneurysms of the bilateral internal carotid arteries (ICAs), which led to a definitive diagnosis. The patient was successfully treated with anticoagulants and antiplatelet agents. The present case study suggests that, for patients with space-occupying lesions of the skull base and symptoms of cranial nerve palsy, the possibility of an ICA dissection should be prioritized during the differential diagnosis.

Introduction

An arterial dissection is a blister-like delamination between the inner and outer walls of a blood vessel, caused initially by a tear that allows blood to enter the inner lining. This results in arterial occlusion, stenosis or aneurysm formation.

Bilateral dissections of the internal carotid artery (ICADs) remain rare, however, ICADs, which were once considered to be extremely rare, are increasingly being recognized as a common cause of stroke in individuals between 35 and 50 years of age due to improvements in diagnostic technologies (1-4). ICADs may be extracranial or intracranial; the latter are usually more severe and exhibit a poorer prognosis. ICADs typically occur in individuals aged between 35 and 50 years, with a slight preponderance among females (2,3,5).

ICADs may occur as a result of trauma, however, the majority appear to occur spontaneously (6). The etiology and pathogenesis of arterial dissections remain largely unknown. However, it has been hypothesized that numerous factors may contribute to their development, including hereditary connective tissue disease (7), Marfan syndrome, Ehlers-Danlos syndrome, hyperhomocysteinemia (8), α -1 antitrypsin deficiency (9), respiratory tract infections (10), migraines (11), hypertension (12) and the use of contraceptives (13).

Common clinical manifestations of ICADs include headache or facial pain, which is often unilateral (6,14), cerebral ischemic events (6,15), dizziness and Horner's syndrome (15). Notably, >10% of patients with a spontaneous ICAD develop cranial nerve palsy (6,14) where the trigeminal, facial, oculomotor, trochlear and abducens nerves (6,14,15), as well as the end-group cranial nerve (6,16) may be affected. In patients exhibiting end-group cranial nerve palsy, which occurs in 3-12% of ICAD cases (17), Collet-Sicard syndrome and Villaret's syndrome often develop (18,19). The hypoglossal nerve is one of the most frequently affected cranial nerves in ICAD (6,14,16,20). Hypoglossal nerve palsy may result from the compression of surrounding structures by the dilated dissecting aneurysm or involvement of blood vessels that supply blood to the cranial nerves.

In the majority of ICADs, local and ischemic manifestations occur within days or weeks following the onset of the first symptoms (21,22). However, persistent dissecting aneurysms that cause thromboembolic complications years after the initial event are rare and thus, may lead to misdiagnosis and mismanagement.

ICADs are usually diagnosed following the detection of an aortic intramural haematoma on computed tomography (CT) or magnetic resonance imaging examination (23). At present, treatments include anticoagulation, stent implantation and carotid artery ligation. Patient outcome is dependent on the severity of the disease. A number of studies have reported a good prognosis in 70-90% of patients without stroke sequelae (24-26).

In this study, a case of old dissecting aneurysms of the bilateral ICAs presenting as space-occupying lesions with skull base erosion and hypoglossal nerve palsy, which subsequently lead to diagnostic confusion, is presented. The aim of this case report was to highlight that in patients with space-

Correspondence to: Dr Yi-Ping Li, Department of Neurosurgery, First Hospital of Jilin University, 71 Xinmin Street, Changchun, Jilin 130021, P.R. China E-mail: liyiping5931@gmail.com

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Figure 1. Computed tomography revealing extensive erosion of the skull base, as indicated by an area of equal density with unclear borders in the lesions and irregular high density in the periphery.



Figure 2. Histological analysis indicating thrombosis of the lesion. Gray areas admixed with dark red areas are evident, and aggregation of platelets, along with entrapped red blood cells are visible (hematoxylin and eosin staining; magnification, x200).

occupying lesions of the skull base and symptoms of cranial nerve palsy, the possibility of ICAD must be considered in the differential diagnosis. Written informed consent was obtained from the patient.

Case report

In July 2006, a 45-year-old female was admitted to the First Hospital of Jilin University (Changchun, China) with a five-year history of intermittent headaches and a two-month history of deviation of the tongue toward the left upon protrusion. Upon admission, a neurological examination revealed no positive signs of nervous system dysfunction, except for the tongue deviation.

A CT scan of the skull base revealed patchy erosion of the base of the middle cranial fossa, the right clivus, the petrous apex of the temporal bone and the right sphenoid bone, and structural disappearance of the wing of the right sphenoid bone (Fig. 1).

The patient received general anesthesia in order to undergo a biopsy of the lesion via an endonasal transsphenoidal approach. Pathological analysis of the lesion revealed the aggregation of platelets and entrapped red blood cells were visible, which suggested a thrombosis (Fig. 2). Subsequently, the patient was diagnosed with a dissecting aneurysm of the bilateral internal carotid arteries.

The patient was administered anticoagulant (100 mg aspirin, once a day) and antiplatelet (150 mg Plavix, once a day) agents. Subsequent to two weeks of therapy, the symptoms of hypoglossal nerve paralysis and headaches had been completely resolved. Despite this, the patient succumbed to epistaxis six months after the surgery.

Discussion

Bone erosion and soft-tissue shadows in the skull base were the major CT findings in the present study. As a result, the patient was pre-operatively misdiagnosed with skull base tumors. However, the pathological analysis of the lesion and a magnetic resonance angiogram examination resulted in a final diagnosis of dissecting aneurysms of the bilateral internal carotid arteries.

In recent years, ICADs have been increasingly recognized to be a common cause of stroke in individuals between 35 and 50 years of age. A stroke typically occurs within the early days or weeks following the onset of symptoms (21,22). In the present case, the patient had experienced intermittent headaches, which had lasted for five years. In addition, the presence of bone erosion in the patient indicated that the lesion had been present for a number of years.

Cranial nerve palsy occurs in >10% of patients with spontaneous dissection of the internal carotid artery (6,14), and the lower cranial nerves IX-XII are most commonly affected, particularly the hypoglossal nerve (6,16). Previous studies have reported the involvement of various combinations of nerves (6,15), however, at present, there are a limited number of studies that describe ICADs manifesting as space-occupying lesions with skull base erosion (21). The present study suggests that, for patients with single or multiple space-occupying lesions of the skull base and end-group cranial nerve palsy, a differential diagnosis should be established from malignant skull base tumors, schwannoma, soft meningioma, infectious diseases and spontaneous diseases (9).

The management of ICA dissections remains controversial. Despite the lack of evidence from randomized clinical trials, it is generally recommended that 3-6 months of anticoagulant therapy should be administered to prevent the occurrence of cerebral ischemia, local compression or rupture (27,28). Carotid artery stenting has also been used to manage ICA dissections, however, there is no definitive evidence that stent placement is beneficial (23). Conservative treatment is a good choice for patients without ischemic complications as a result of dissecting aneurysms, which has been shown to exhibit significant efficacy in relieving hypoglossal nerve palsy (6,14,16,29). In the present study, subsequent to considering the advantages and disadvantages of each potential treatment, anticoagulant and antiplatelet agents were administered to the patient in order to prevent further thrombosis. The patient initially recovered well with no pathological recurrence, however, succumbed to epistaxis six months after surgery.

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