CASE REPORTS/CLINICAL VIGNETTES

Spontaneous Dissection of Internal Carotid Artery Masquerading as Angioedema

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Spontaneous dissection of the internal carotid artery usually presents with stroke-like symptoms secondary to ischemia in its vascular territory, as well as local signs and symptoms, which may include head, face or neck pain, Horner's syndrome, pulsatile tinnitus, and cranial nerve palsies. We report a case of a 44-year-old healthy white male who presented with tongue swelling mimicking angioedema as an unusual manifestation of spontaneous dissection of the internal carotid artery. Two weeks after the initial presentation, the patient returned with similar symptoms and slurred speech. Upon physical examination, he was noted to have isolated left-sided hypoglossal nerve palsy. Subsequent diagnostic imaging revealed segmental narrowing of the left internal carotid artery. The appearance was consistent with the presence of a spontaneous internal carotid artery dissection with associated pseudoaneurysm formation.

KEY WORDS: dissection; internal carotid artery; angioedema.

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INTRODUCTION

Spontaneous dissection of the internal carotid artery (ICA) usually presents with stroke-like symptoms secondary to ischemia in its vascular territory. Additional local signs and symptoms may include head, face or neck pain, Horner's syndrome, pulsatile tinnitus, and cranial nerve palsies. The objective of our manuscript is to report an unusual presentation of ICA dissection as tongue swelling mimicking angioedema. Our review of the literature failed to reveal another case with similar findings. In this report, we aim to describe this unusual presentation of ICA dissection and also discuss common clinical manifestations. An isolated hypoglossal nerve palsy and radiographic imaging directed our diagnosis and treatment. ICA dissection should be considered in the differential diagnosis of patients who present with isolated hypoglossal nerve palsies.

CASE REPORT

A 44-year-old white male, without any significant past medical history, presented with swelling of the tongue, dyspnea and chest tightness for two days. The patient denied any history of trauma, chiropractic manipulation, use of medications, or exposure to any known allergen. On initial presentation, his physical examination revealed apparent swelling of his tongue. The remainder of his exam was normal. Routine laboratory tests were unremarkable. A lateral roentgenogram of the neck suggested narrowing of the airway in the hypopharyngeal area. A diagnosis of angioedema with possible laryngeal edema was considered. The patient was electively intubated for airway protection and placed on mechanical ventilation. He was treated with high dose intravenous methylprednisolone and racemic epinephrine to decrease the laryngeal edema. He was also given cetirizine and cimetidine for their anti-histaminergic properties. Evaluation for hereditary angioedema, including serum assays for C3 and C4 complement levels as well as C1-esterase inhibitor were all found to be normal. He was discharged home after resolution of his symptoms.

Two weeks later, the patient returned to the emergency room with similar symptoms as well as slurred speech. He denied trauma or any form of chiropractic manipulation since his discharge from the hospital. On physical examination, the tongue was mildly swollen and was deviated to the left side, suggestive of left hypoglossal nerve palsy. The remainder of his examination including the cranial nerves, motor and sensory functions were all normal. Magnetic resonance imaging (MRI) of the head and neck revealed "fluid signal process intimate to the internal carotid artery" and in close proximity to the course of the hypoglossal nerve. A subsequent magnetic resonance angiography (MRA) of the neck followed by conventional angiography revealed segmental narrowing of the left ICA from the level of the C2 vertebrae extending into the petrous segment of the vessel. The middle segment of the narrowed left ICA was associated with an aneurysmal outpouching measuring 1.1 cm in length and 0.8 cm in diameter, see Figure 1. These radiographic findings were consistent with a spontaneous ICA dissection with associated pseudoaneurysm formation.

DISCUSSION

Spontaneous internal carotid artery dissections are uncommon. In community-based studies in the United States and France, the annual incidence of spontaneous ICA dissections ranged from 2.5 to 3 per 100,000^{1,2}. Spontaneous dissections of the carotid and vertebral arteries affect all age groups,

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Figure 1. Carotid Angiogram demonstrating segmental narrowing of the left internal carotid artery with associated pseudoaneurysmal outpouching. Appearance is consistent with presence of an internal carotid artery dissection.

including children; however, ICA dissections present most commonly in the fifth decade of life³⁻⁶. At presentation, on average, women are five years younger than men, although there is no gender predilection³. Patients with spontaneous dissection of the carotid artery are believed to have an underlying structural defect of the arterial wall, albeit the exact type of arteriopathy remains elusive in most cases. Among the heritable connective tissue disorders associated with an increased risk of spontaneous ICA dissection, Ehlers-Danlos syndrome type IV is most common⁷. Others include Marfan's syndrome, autosomal dominant polycystic kidney disease, and osteogenesis imperfecta type I^{7,8}. Minor events such as hyperextension or rotation of the neck while practicing yoga, painting a ceiling, coughing, vomiting, or sneezing have all been reported to precipitate ICA dissection. Chiropractic manipulation of the neck has also been associated with ICA dissection¹⁵. Additionally, infections such as purulent tonsillitis, purulent and nonpurulent pharyngitis, otitis media and sinusitis have also been described as potential risk factors¹⁶. It may also occur during intubations for general anesthesia or cardiopulmonary resuscitation^{4,9-14}. Mechanical ventilation as a direct cause of ICA dissection has not been reported in the literature. In our patient, we do not think the procedure of intubation could have been responsible for ICA dissection as it was done electively and without the use of any significant physical force. Moreover, it was performed by an experienced physician under fiberoptic guidance. Most cases of ICA dissections present within hours; however, some develop over weeks as evolving symptoms leading patients to seek medical attention¹⁷.

Symptoms of ICA dissection include headache, anterior neck pain, scalp tenderness, transient ischemic attack, cerebral infarction, amaurosis fugax, cranial nerve palsies, tinnitus and Horner's syndrome¹⁸⁻²⁰. Sturzenegger et al.²¹ reported

44 patients with spontaneous ICA dissection with symptoms including unilateral headache (68%), transient ischemic attack (20%) and cerebral infarction (9%).

Ipsilateral cranial nerve (CN) palsies have been reported in association with ICA dissection²². This case report serves to remind us to consider the diagnosis of ICA dissection in patients presenting with isolated hypoglossal nerve palsy. Mokri et al. described a prevalence of 12% of CN palsies in their study of 190 adult patients with spontaneous extracranial ICA dissection²⁰. Those with lower CN palsies (5.2%) included variable involvement of the hypoglossal nerve along with CN IX, X and XI, and only one case with an isolated hypoglossal nerve palsy. More common CN palsies included CN V (3.7%) and CN III (3.6%)²⁰. The mechanism of cranial nerve palsy in ICA dissection has been theorized to be secondary to compression or stretching of the nerve by the expanded artery. An alternative mechanism is likely interruption of the nutrient vessels supplying the nerve²⁰.

MRI and MRA have a sensitivity and specificity of >90% and are increasingly becoming the diagnostic test of choice for extracranial ICA dissection. Cervical and transcranial Doppler ultrasound imaging can be used for rapid diagnosis in an emergent setting²³. However, carotid and cerebral angiography remain the gold standard for diagnosis. A tapered stenotic segment, known as the "string sign", is the most common positive finding, noted in 32 out of 46 (70%) patients with ICA dissection by Mokri et al.²⁴.

Systemic anticoagulation remains the treatment of choice for ICA dissections. Initial therapy includes intravenous heparin with concomitant institution of oral warfarin for conversion to long term oral therapy. The target international normalized ratio is maintained between 1.5 to 2.5²⁵⁻²⁷. In patients with progressive neurologic deficits, endovascular stenting and balloon dilatation are successful alternatives to surgical therapy, which if needed consists of ligation of the carotid artery combined with an in-situ or extracranial-to-intracranial bypass^{28,29}. Spontaneous ICA dissections usually have a favorable prognosis, with approximately 90% of cases resolving without serious sequelae. Spontaneous recanalization of the ICA is reported in over two thirds of patients with a recurrence rate of less than 10%²⁶. Our patient was treated with heparin and warfarin with resolution of his symptoms, supporting his diagnosis of ICA dissection, although his initial presentation and response to steroids cannot be explained. This diagnosis was initially overlooked as he presented with symptoms suggestive of a more common condition, specifically angioedema.

CONCLUSION

Our case highlights unique clinical findings including an initial presentation with apparent tongue swelling mimicking angioedema in a patient with spontaneous ICA dissection. A heightened clinical index of suspicion is needed for the diagnosis of this disease entity especially in patients with subtle manifestations such as our patient. ICA dissection should be considered in the differential diagnosis of patients presenting with isolated hypoglossal nerve palsy, although most cases present with local signs and ischemic stroke-like symptoms. With increasing availability and sensitivity of MRI and MRA, a larger number of patients with ICA dissection will be detected early

in the course of the disease process. Fortunately, the prognosis remains favorable in most cases.

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