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Internal Carotid Artery Dissection Associated with An Elongated Hyoid Bone in A Patient with Vascular Ehlers-Danlos Syndrome

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Keywords:

Ehlers-Danlos Syndrome; Hereditary collagen disorders; Arterial dissection

1. Abstract

This mini-review presents a compelling case study of a young individual who experienced a transient ischemic attack (TIA) linked to a dissection of the internal carotid artery, ultimately diagnosed with vascular Ehlers-Danlos syndrome (vEDS). The patient's symptoms began after heavy lifting and sudden neck movements, leading to significant neck pain, dizziness, and sensory loss, raising concerns for acute neurological events.

2. Introduction

Structural damage of the internal carotid artery caused by the hyoid bone is an extremely rare phenomenon. An elongated hyoid bone can entrap, hook, or protrude into the internal carotid artery (ICA) which can later cause irregularities of the arterial wall. This phenomenon has been implicated in the pathogenesis of thrombi formation which may lead to cerebral thromboembolic events [1]. We present a case of a young patient who presented to the emergency department with a transient ischemic attack (TIA) due to an internal carotid artery (ICA) dissection which was found to be a direct result of the close contact between an elongated hyoid bone and the artery. Molecular genetic testing was later performed on the patient which detected a heterozygous COL3A1 pathogenic genotype, confirming the diagnosis of vascular Ehlers-Danlos syndrome (vEDS). This case highlights the importance of assessing the possibility of a carotid dissection through an appropriate clinical history and a six-vessel angiogram for its diagnosis.

3. Key Findings

Imaging studies revealed a dissection of the right common carotid artery, confirmed through a six-vessel cerebral angiogram. Genetic testing identified a mutation in the COL3A1 gene, characteristic of vEDS, suggesting a hereditary connective tissue disorder contributing to the patient's arterial fragility.

4. Discussion

This article highlights the rarity of neurovascular events due to carotid artery damage from cervical anatomy, noting potential mechanisms such as turbulent blood flow during neck movements. The article discusses differential diagnoses, including Eagle syndrome, while emphasizing the unique presentation of TIA in this context. Worldwide only few cases have been published concerning neurovascular events as a result of structural damage to the internal carotid artery caused by cervical bone structures. The spectrum of symptoms reported in literature ranges from ipsilateral ischemic cerebrovascular event to eagle syndrome [2-5]. To the best of our knowledge, this is the first report of a transient ischemic attack in this scenario. In this case, the patient's symptoms may have arisen from changes in the blood vessels caused by structural damage due to repeated local trauma from the close proximity of the hyoid bone to the internal carotid artery (ICA). During the angiography, it was observed that certain neck movements, such as rotating away from the lesion, could increase turbulence in blood flow, potentially leading to the formation of blood clots. Additionally, actions like swallowing and forceful coughing caused the hyoid bone to shift against the lateral wall of the ICA. The contact between the ICA and the hyoid bone was only identified after a CT angiogram was conducted. Initially, a doppler ultrasound of the carotid arteries

revealed a dissection of the ICA, but the underlying cause was unclear. It is crucial to carefully examine various angles and maintain a high suspicion of the structural and positional relationships of the ICA as a potential risk factor for disease. A subsequent cross-sectional CT angiography proved to be an effective imaging technique for identifying the location of the artery dissection and its relationship to important neck structures. Though general treatment guidelines for carotid dissections are available, recommendations about dealing with an elongated hyoid bone cannot be given at the moment. Previous published cases have also included the resection of the greater horn of the hyoid bone as part of their treatment plan,4,5 as well as resection of the injured segment of the ICA. A study in Brazil examined the irregularity of the cell wall of the injured segment where microscopic changes, including intimal and endothelial damage, were observed due to the continuous mechanical compression of the ICA by the hyoid bone.3 However our patient remained without impingement after the resection of the posterior horn was performed, therefore endarterectomy was found to be unnecessary. After confirming the diagnosis of vEDS, he did note a history of spontaneous bruising throughout his life. After eliciting a detailed medical profile, we learned he had a number of spontaneous deaths in young relatives due to a uterine hemorrhage, an arterial rupture and an acute peritonitis due to perforation of a digestive tract. On examination his face had large eyes, thin nose and lips, a small chin and lobeless ears were observed (Figure 10). Skin elasticity was normal. Distal interphalangeal joints and shoulders were hyperextensible with multiple movements (Figure 10). Metacarpophalangeal joints hyperextended > 90°, hyperflexion of wrists approached 140°, and his knees achieved hyperextension > 15°. Transthoracic echocardiography did not show mitral valve prolapse or other abnormalities. No causal treatment for vEDS is available, and the current treatment consists mainly of the prevention of complications, as well as symptomatic treatment, including surgery and catheterization. Thus, close outpatient follow-up and appropriate guidance to the patient and family should be provided.6 Following treatment with dual antiplatelet therapy and surgical intervention to alleviate external compression on the internal carotid artery, the patient showed remarkable recovery, with no residual symptoms at follow-up.

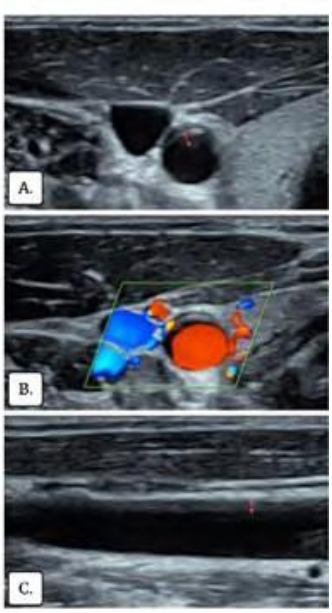

Figure 1: Doppler ultrasound showing dissection of right the common carotid artery. **A.** On transverse view (*arrow*). **B.** Color doppler flow image with careful adjustment of colour gain detected the intimal flap. **C.** On longitudinal view (*arrow*).

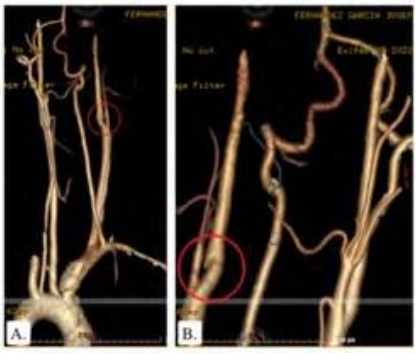

Figure 2: Three-dimensional reconstruction of an angiotomography analysis of the carotid arteries showing the dissection of the right ICA (*red circle*) (**A** and **B**).

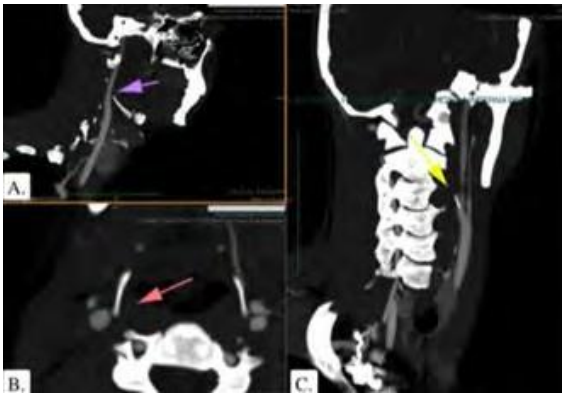

Figure 3: CTA demonstrated a close contact between the enlarged right posterior horn of the hyoid bone and the right ICA. **A** and **C**, sagittal view. **B.** Axial cut of the CT scan. (*Purple arrow*: posterior horn of the hyoid bone, *red arrow*: relationship between ICA and ECA, *yellow arrow*: communication between the hyoid bone and ICA).

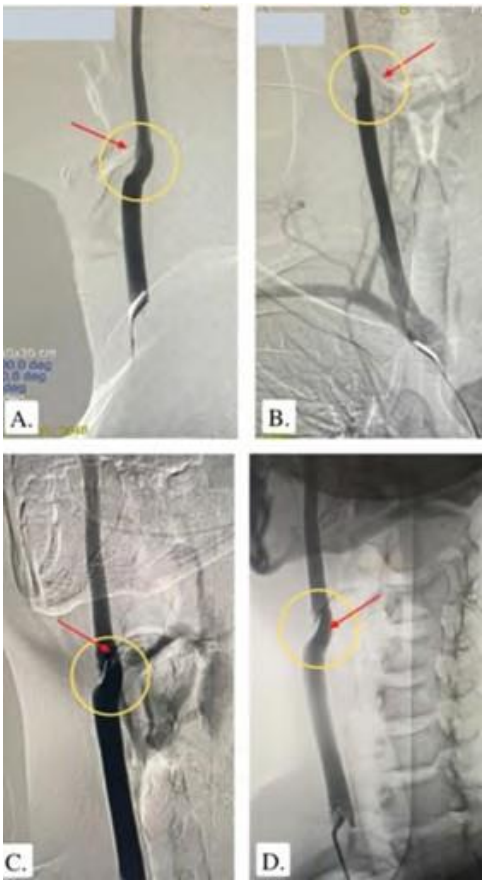

Figure 4: DSA images reveal dissection of the right ICA (*encircled in yellow*) and its close contact with the hyoid bone (*red arrow*). **A.** Lateral projection. **B.** Anteroposterior projection. **C** and **D.** Oblique projection.

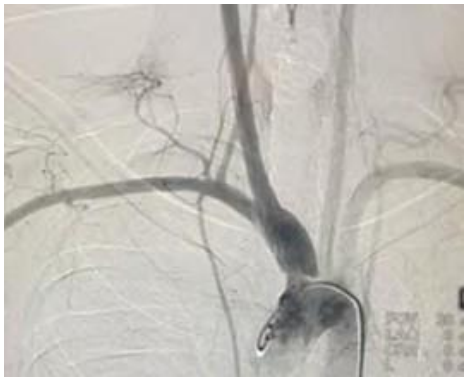

Figure 5: DSA images in an anteroposterior projection reveal dissection involving the origin of theright brachiocephalic artery.

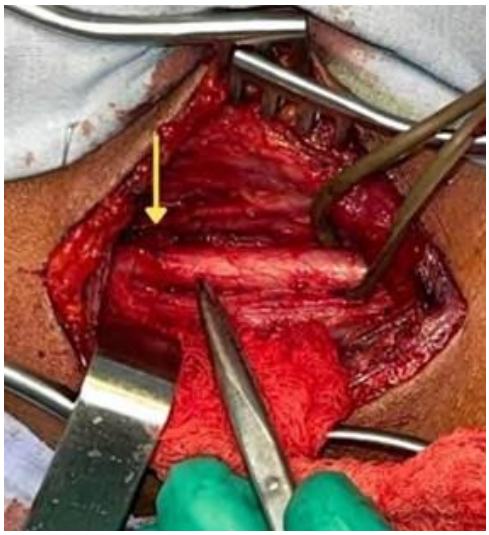

Figure 6: Intraoperative exposure of the internal carotid artery after resection of the posterior horn of the elongated hyoid bone showing no sign of impingement.

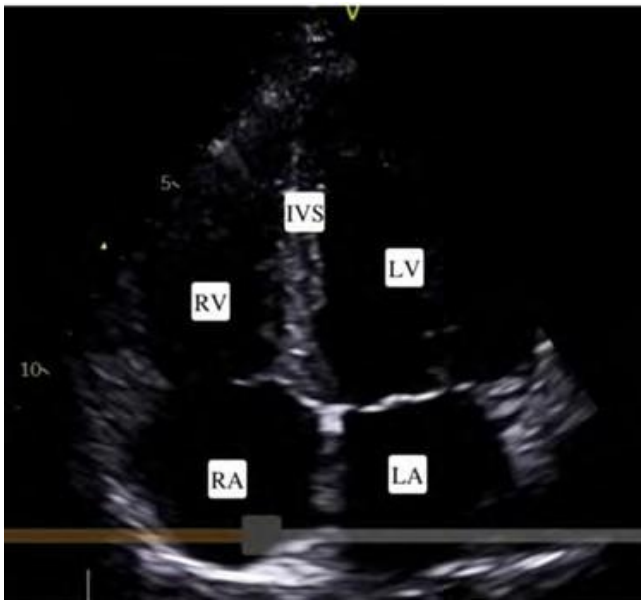

Figure 7. An apical 4 chamber view obtained with a phased array transducer in the left fifthintercostal space, with the probe marker corresponding to the patient's right. Abbreviations: RA (right atrium), LA (left atrium), RV (right ventricle), LV (left ventricle) and IVS (interventricular septum).

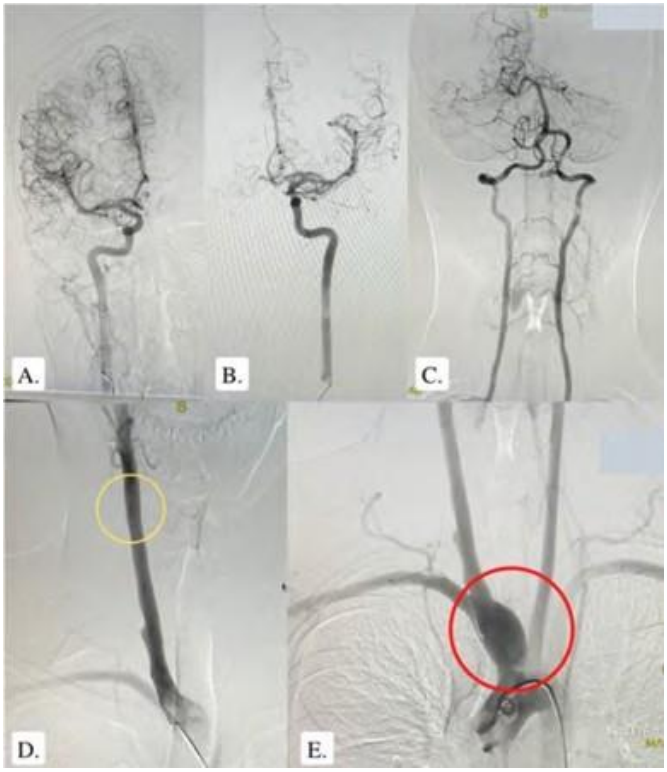

Figure 8: DSA images reveal resolution of right ICA (*encircled in yellow*) and an aneurysmal dilatation was observed on the right proximal brachiocephalic trunk (*encircled in red*). **A.** Anteroposterior projection of a selective injection of right ICA showing patency of the right anterior communicating artery with retrograde filling of the left ICA and terminal branches. **B.** Anteroposterior projection of a selective injection of the left ICA. **C.** Anteroposterior projection of the posterior circulation. **D.** Anteroposterior projection of the right CCA. **E.** Anteroposterior projection reveal dissection involving the origin of the right brachiocephalic artery.

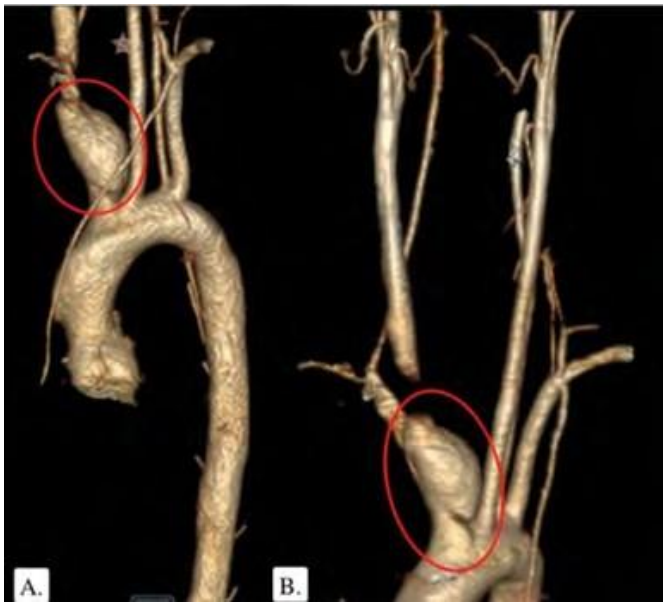

Figure 9: Three-dimensional reconstruction of an angiotomography analysis of the carotid arteries showing the dissection of the right brachiocephalic trunk associated with a fusiform aneurysmal dilatation with the following dimensions: 32x 18.8 mm (*red circle*) (**A** and **B**).

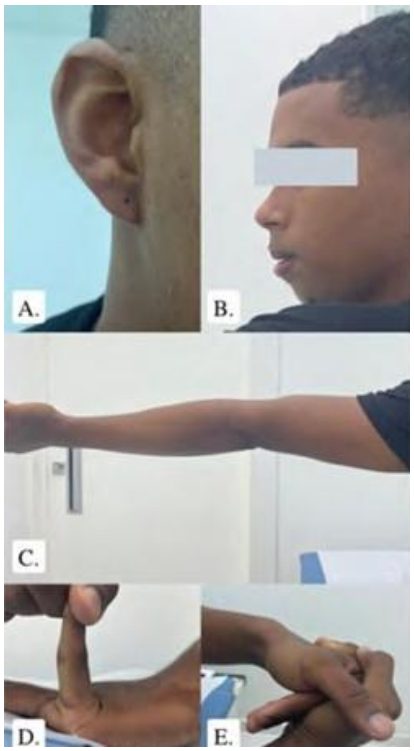

Figure 10. Patient clinical features: (**A.**) lobeless ears, (**B.**) long thin nose and lips with a triangular-shaped face and a small chin (**C.**) hypersxtension of elbows > 10°, (**D.**) passive extension of just the little finger beyond 90° with the forearm flat on a table and (**E.**) passive flexion of the metacarpophalangeal joints.

5. Conclusion

Identifying the risk of a neurovascular event related to cervical structures is essential for effective management and prevention of future occurrences. A comprehensive review of a six-vessel cerebral digital subtraction angiography is particularly important, especially if the patient can perform movements like head rotation and swallowing.

This article highlights the significant risk of dissection and ischemic stroke associated with cervical bones near the internal carotid artery (ICA), particularly in otherwise healthy young patients who experience recurrent strokes or transient ischemic attacks (TIAs) of unknown origin. Molecular genetic testing confirmed a diagnosis of vascular Ehlers-Danlos syndrome (vEDS), revealing that the patient is heterozygous for the COL3A1 c.1379_1406delinsA p. Ala460_Ser469delinsGlu mutation. The patient is currently receiving double antiplatelet therapy following an arterial dissection and has not experienced any new vascular events in the past year. This diagnosis requires a high degree of clinical awareness and proactive management to prevent serious consequences.

This case contributes valuable insights to the limited literature on cervical vascular complications and highlights the need for thorough genetic evaluation in similar cases. The findings advocate for awareness of atypical presentations of common vascular issues in young patients, emphasizing the interplay between structural anomalies and ischemic events.

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