A Rare Variant of Eagle's Syndrome

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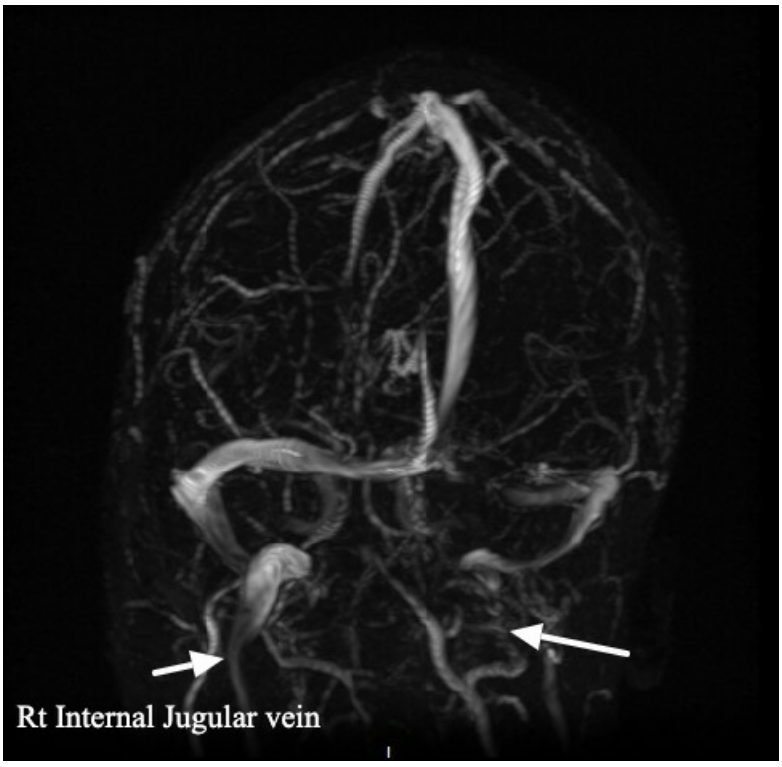
Introduction: Eagle’s syndrome(ES) is a rare condition associated with abnormality/ossification of styloid process. We report a case of young male who was found to have internal jugular venous stenosis (IVJS) due to extrinsic compression by styloid process. Case Report: 25-years-old male with no significant history presented with left-sided neck and jaw pain with associated left-sided headache. He also reported photophobia and chronic posterior neck pain. Physical examination was remarkable for left sided neck and jawline tenderness. Laboratory findings were within normal limits. Possibility of subarachnoid hemorrhage and CNS infections was ruled out with normal CSF and no aneurysm on CT-Angiogram (CTA). CTA neck revealed reflux of contrast into left jugular vein with abrupt cut off at the proximal aspect and complete opacification of the right jugular vein resulting in asymmetric opacification. Patient was started on therapeutic enoxaparin for possible venous thrombosis which was ruled out by Magnetic Resonance Venography (MRV) Brain which did reveal poor flow related signal within the left internal jugular vein below the skull base, likely relating to compression between the styloid process and C1 vertebral body, with compensatory increased drainage through the left pterygoid and vertebral venous plexuses [Figure 1]. Patient reported resolution of symptoms during admission and was discharged with neurosurgery follow-up for possible decompression or stenting. Discussion: Eagle defined two syndromes associated with an elongated styloid process, classic syndrome and Stylo-carotid syndrome. Classic syndrome is characterized by pain and dysphagia, and carotid variant refers to stenosis or dissection of internal carotid artery caused by compression by styloid process. In both syndromes, styloid process length seems to be the major cause. However, literature describes another very rare variant, Stylo-Jugular variant as in our case. IVJS secondary to compression between elongated styloid process coursing adjacent to the transverse process of C1, causing venous reflux obstruction has been termed stylo-jugular ES. Extrinsic compression of IJV especially by styloid process is one of the important etiologies of IJVS. Ipsilateral pain is usually more significant symptom. Headache, numbness, and dizziness are usually chronic and may be related to the impaired cerebral venous outflow. MRV is preferable to CT Venography in terms of accuracy of diagnosis. The treatment is usually conservative, especially when symptoms are not invalidating and apparently controlled by medical treatment. Jugular stenting has been reported to show benefit. More investigation is warranted to study the symptoms and treatment of this condition.



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