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Eagle syndrome an overlooked cause of internal carotid artery dissection, Case series

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ABSTRACT

Background: Eagle syndrome is a rare condition with a prevalence estimated at 4%, with 0.16% exhibiting symptoms of Eagle's syndrome resulting from an elongated styloid process or a calcified stylohyoid ligament, compressing over nearby vessels and nerves. Symptoms range from simple headache, neck pain, and odynophagia to cerebral ischemia. Since it can be an incidental finding, management is controversial and ranges from conservative treatment to surgical intervention, including internal carotid stenting and styloidectomy.

Clinical presentation: We report two cases of Eagle's syndrome. Case one (Vascular variant): 44 years old presented with left-sided weakness with Magnetic Resonance Imaging and an angiogram of the brain showing a right middle cerebral artery hemispheric infarct with a right internal carotid artery (ICA) dissection and elongated styloid processes more on the left. The patient's condition improved with supportive care. He was treated with aspirin, atorvastatin, and an intensive 16-week rehabilitation program. Case two (Classic variant): 41 years old presented with refractory occipital headache and neck pain without vascular insult. Magnetic resonance of both patients showed a long styloid process.

Conclusions: Eagle's Syndrome is a rare entity and perhaps overlooked as an anatomical variant that ought to be considered a possible etiology in atraumatic ICA dissection in patients with no discernable risk factors. As observed in case one, with atraumatic ICA dissection and unremarkable connective tissue disease work-up, highlighting anatomical variant, as an etiology to his ischemic stroke. Styloidectomy should be considered to avert risk of recurrent IC dissections in such patients with atraumatic IC dissection and classic eagle's syndrome with refractory orofacial pain syndromes exacerbated by head movement or yawning.

1. Introduction

In1937, Eagle's syndrome was first described as a cluster of symptoms implicated by the compression of an elongated or ossified styloid process (osteoligamentous malformation) onto nearby anatomical structures [1]. In literature,afew variants have been described, with the most commonly observed are two; the classical variant, characterized by odynophagia owing to glossopharyngeal irritation, and the vascular Eagle's syndrome (Carotid or Jugular variant) that presents with headaches, and cerebral ischemia relative to recurrent head and neck movements [1–5].

The prevalence of elongated styloid process among the general population is estimated at 4%, with 0.16% exhibiting symptoms of Eagle's syndrome [5]. Owing to the debate on the implicated pathophysiology of such entity from a possible impingement of pharyngeal apparatus and traversing neurovascular structures, such as

Abbreviations: ICA, Internal carotid artery; ICAD, Internal carotid artery dissection; IV, intravenous; OPG, orthopantomogram; SPRD, Styloid process related dissection; ED, emergency department; MRI, magnetic resonance imaging; MIP, Maximum intensity projection; DTR, deep tendon reflexes; ADLs, activities of daily living; 3D CT, Three-dimensional computed tomography; FLAIR, fluid attenuated inversion recovery; TOF, Time of flight; DWI, Diffusion weighted imaging; ADC, Apparent diffusion coefficient; ANA, autoimmune screen (anti-nuclear antibody); ANCA, antineutrophil cytoplasmic antibody.

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glossopharyngeal or extracranial Internal carotid artery (ICA), vascular variant prevalence ranges from (4–32.9%); however, it is postulated to be underestimated as an etiology of ICA dissection and cerebral ischemia [5].We herein report two rare cases of Eagle's syndrome with a clinical manifestation of the classical variant and one with a vascular variant that resulted in cerebral ischemia due to ICA dissection.

2. Case presentation

Case 1. A 44-year-old right-handed nonsmoker male with a past medical history of uncontrolled hypertension was brought to the emergency department (ED) within one hour of the sudden onset left sided weakness. Patient symptoms started while driving with right-sided aching paroxysmal, gradually worsening headache of a severity of 8/10 accompanied by sudden onset left-sided hemiparesis, dysarthria, and hemi-hypoesthesia. Significantly, he has been exhibiting nine months history of progressive right-sided paroxysms of cervical-facial pain described as aching with superimposed shooting electrical sensation aggravated by neck movements for which he never sought medical attention.

Upon hospital arrival, His vital signs showed a temperature of 36.7C, Heart Rate of 94 bpm, respiratory rate of 23 breaths/min, blood pressure of 163/108 mmHg, and oxygen saturation of 99 % on room air. He appeared drowsy with no clear extrapyramidal signs. The National institute stroke scale (NIHSS) of 18 with right, forced gaze deviation, left homonymous hemianopia, flattening of the left nasal fold, left-sided dense hemiplegia and hypoesthesia, hemispatial neglect with tactile inattention. However, he retained the ability to follow written and verbal commands, suggestive of intact comprehension and language function. A multimodal CT head was performed. (Fig. 1 A). The plain CT head did not reveal any intracranial hemorrhage and no established hypodensities. Hence intravenous (IV) tissue plasminogen activator (tPA) 0.9 mg/kg was given after written informed consent.

One hour post thrombolysis NIHSS has improved to 4 with residual left-sided drift in upper and lower limbs, decreased sensation, and subtle dysarthria. CT angiogram showed a flame sign with occlusion of the right ICA approximately 1.2 cm distal to the carotid bifurcation, with no opacification seen until the supraclinoid ICA (Fig. 1E), suggestive of ICA dissection. Partial opacification of the M1 segment of the right MCA with a filling defect is seen in the distal M1 (Fig. 1D).

Coronal CT of the head, maximum intensity projection (MIP) reconstructed images bony window algorithm showing elongated styloid processes bilaterally thicker and longer on the right side (>3cm) suggestive of Eagle's syndrome (Fig. 1B).

Due to the rapid improvement of the NIHSS with tPA, the interventional radiology and stroke teams decided not to intervene endovascularly, owing to the potential additional risk from possible ICA dissection. Twenty-four hours post-tPA, the patient was noted to have left-sided distal rhythmic writhing movements suggestive of athetosis with residual left upper limb drift and decreased pin-prick sensation likely due to thalamic infarction. Follow-up CT head 24-hour post-tPA; showed no evidence of acute intra-axial or extra-axial hemorrhage with multifocal hypodensities noted in the right front-parietal-temporal lobe representing established right MCA territory infarction. (Fig. 2A–D). Secondary stroke prevention with, Aspirin 100 mg and Atorvastatin 40 mg daily per orally was started.

Forty-eight hours post thrombolysis, the patient developed worsening of the pre-existing symptoms with right forced gaze deviation and left-sided dense hemiplegia with a significant increase in the NIHSS to 21. Urgent CT head showed no evidence of acute intracranial hemorrhage. A large hypodense area is seen in the right frontoparietal cortical and subcortical regions and basal ganglia, denoting a recently established infarct which appeared larger compared to the initial study, suggestive of a further embolic phenomenon. The Magnetic Resonance Imaging (MRI) of the head showed multiple focal right cerebral parenchymal areas of diffusion restriction are seen involving the frontal, temporal, and parieto-occipital region and the right basal ganglia. The

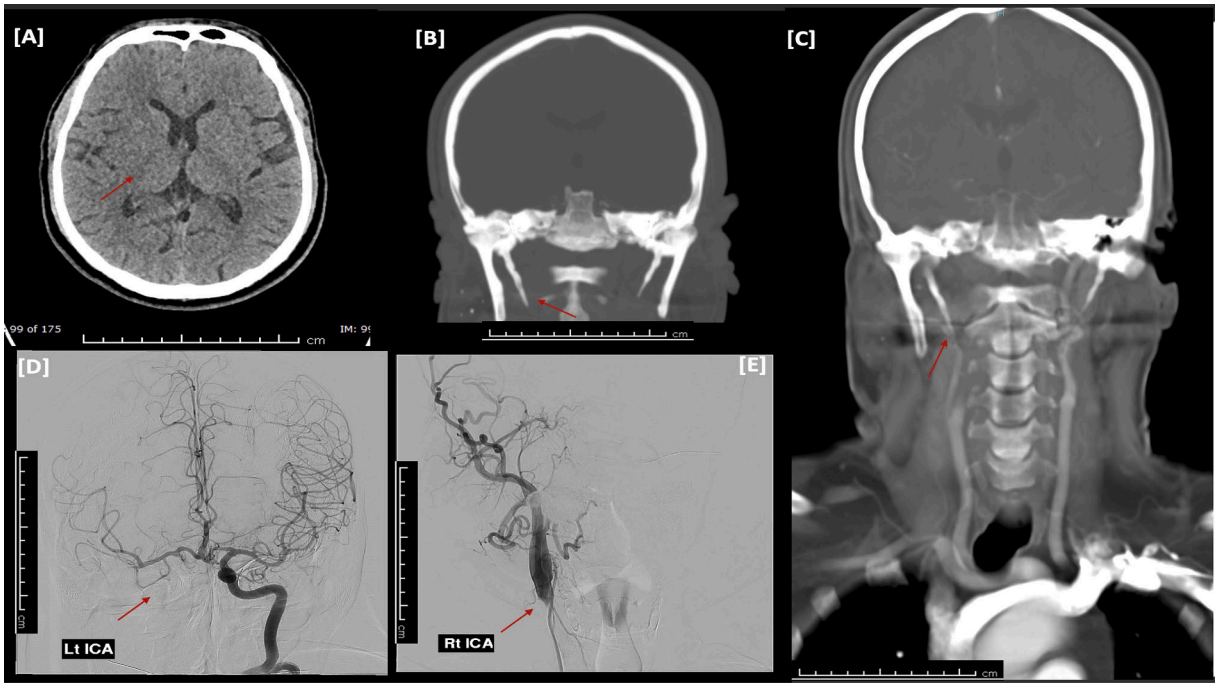

Fig. 1. [A-E] Case one with right ICA dissection and elongated styloid processes. [A] Axial Non-contrast CT brain at the level of the basal ganglia and internal capsules showing subtle obscuration of the borders of the right lentiform nucleus raising the possibility of hyperacute ischemic insult along the right MCA territory. [B] Coronal MIP reconstructed images bony window algorithm showing elongated styloid processes bilaterally thicker and longer on the right side (>3cm). [C] Coronal reconstructed MIP CT angiogram showing the right elongated styloid process reaching the level of the occluded proximal right ICA at the level of the transverse process of C2 vertebra. [D and E] Conventional cerebral angiogram showing total occlusion with absent flow along the right internal carotid artery with refilling of its terminal cerebral branches from the contralateral left internal carotid artery injection.

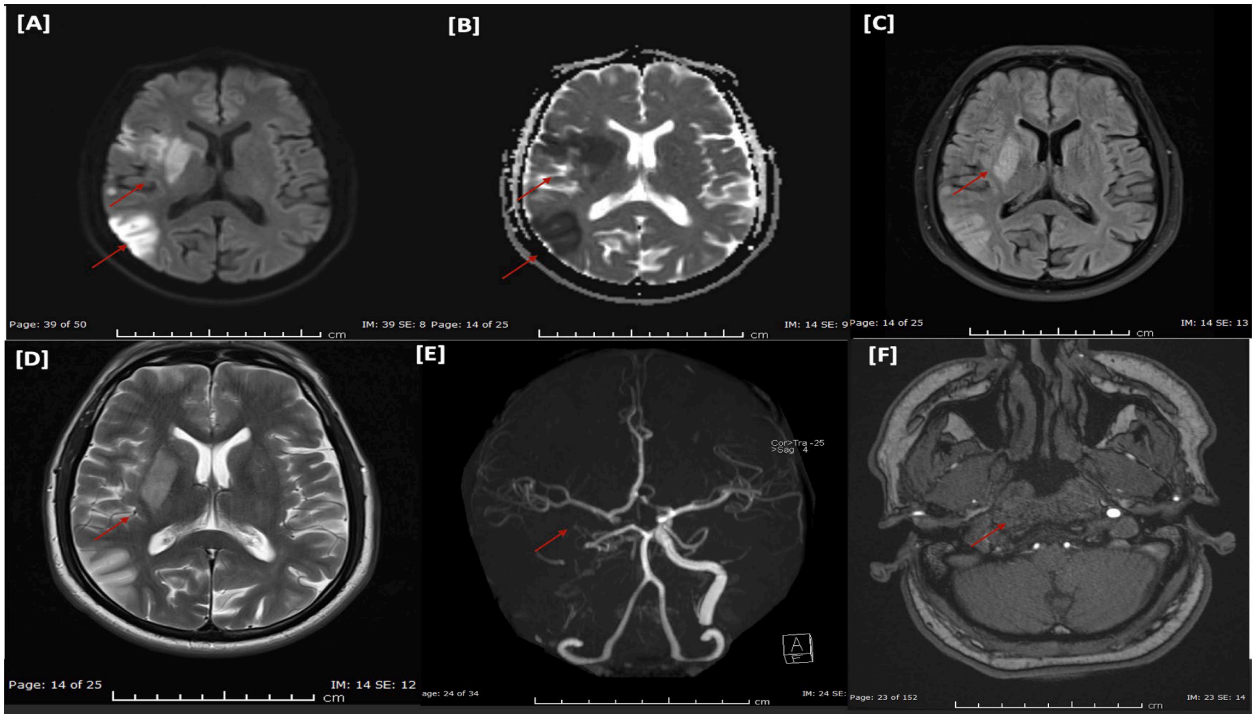

Fig. 2. [A–E] MRI /MRA head – Case one with right ICA dissection and elongated styloid processes. [A and B] Axial DWI and ADC map images of the brain showing recent ischemic infarctions along the right MCA territory in the right temporal and parieto-occipital region involving the right lentiform nucleus. [C and D] Axial FLAIR and T2 WI showing corresponding hyperintensities. [E and F] 3D TOF MRA of the intracranial arteries showed absent flow in the intracranial course of the right ICA while preserved flow noted along the right ACA and MCA.

MR angiogram of the head and neck showed absent flow along the intracranial course of the right internal carotid artery.

Further stroke work up normal sinus rhythm left ventricular hypertrophy and left axis deviation on electrocardiogram (ECG). no evidence of atrial fibrillation on 48-hour Holter monitoring and no evidence of patent foramen ovale (PFO), valvular abnormalities, or ventricular thrombus on echocardiography. Initial labs showed an HbA1c of 5.8%, an LDL level of 108, total cholesterol of 201, a low Vitamin B12 level of 91.5 pmol/L, and an elevated Homocysteine level of 49.9 umol/L where observed and were treated with cyanocobalamin intramuscular injection, 1000mcg daily for one week followed by weekly maintenance for eight weeks. A working diagnosis of ICA dissection was likely due to right prolonged/calcified styloid process suggestive of vascular Eagle's syndrome diagnosis.

The patient's management encompassed a multidisciplinary approach. Over a few days, he was more alert with persistent deficits on the NIHSS of 20. He was transferred to the rehabilitation center to undergo physical therapy. He was maintained on aspirin 100 mg daily per orally and atorvastatin 40 mg daily per orally and was referred to the Head and Neck Surgery outpatient clinic for possible styloidectomy to avert the risk of recurrent dissection in the future.

Case 2. A 41-year-old gentleman right-handed nonsmoker male with a past medical history of dyslipidemia, and multiple episodes of palpitation due to premature ventricular contracture (PVC). He presented to ED with episodes of palpitations, gradual onset occipital headaches of 6 months duration aching in nature, lasting on an average for 4 h with a frequency of 4–5 times per month associated with ear block sensation, numbness in the upper/lower limbs, and pre-syncope.

His vital signs upon arrival to Hospital ED were BP of 134/75, PR 98, he was afebrile maintaining oxygen saturation on room air. His initial blood workup showed a normal complete hemogram, renal & liver functions, and coagulation profile. He had serum levels of cholesterol, LDL, HDL, and triglyceride of 6.1, 4.2, 0.7, and 2.5 mmol/L respectively. ECG this time was unremarkable. Echocardiography showed an ejection fraction of 56 %, with a normal four heart chambers. A Holter tape showed ventricular ectopic beats at a rate of 1.7%. Computed tomography of the brain (Fig. 1) was essentially normal. Magnetic Resonance Imaging (MRI) with contrast and venogram (Fig. 2) showed normal brain parenchyma in the pre and post-contrast sequences, normal venogram with evidence of bilateral impingement of the internal jugular veins between the styloid process and C1 transverse process/adjacent muscle complex and multiple posterior intermuscular collaterals carrying venous flow was also noted. The styloid process length was measured to be >3 cm bilaterally (Fig. 3A-B) suggestive of Eagle's syndrome.

Subsequently, carotid duplex ultrasonography ruled out any focal functional luminal stenosis or flow disturbances bilaterally. The patient's management encompassed a multidisciplinary approach, with pain management with nonsteroidal anti-inflammatory drugs (NSAIDs) as needed and he was referred to the maxillofacial team to explore the option of definitive surgical treatment. The patient is scheduled for follow-up after possible surgical intervention to assess overall symptomatology and avert risk of ICA dissection.

3. Discussion

We highlight a rare, yet important syndrome in two cases of vascular and classic Eagle's syndrome, with one manifested a sequela of ICA dissection and hemispheric stroke and the second with the classic variant to further empathize the relevance of prolonged styloid process in atraumatic ICA dissection and considering surgical management to prevent further recurrent dissections which can over-all improve outcomes.

The tentative pathophysiology and mechanism of the elongated styloid process and its ligamentous calcification remains an area for further research, with proposed mechanisms encompassing altered bone homeostasis and developmental anomalies during embryonic

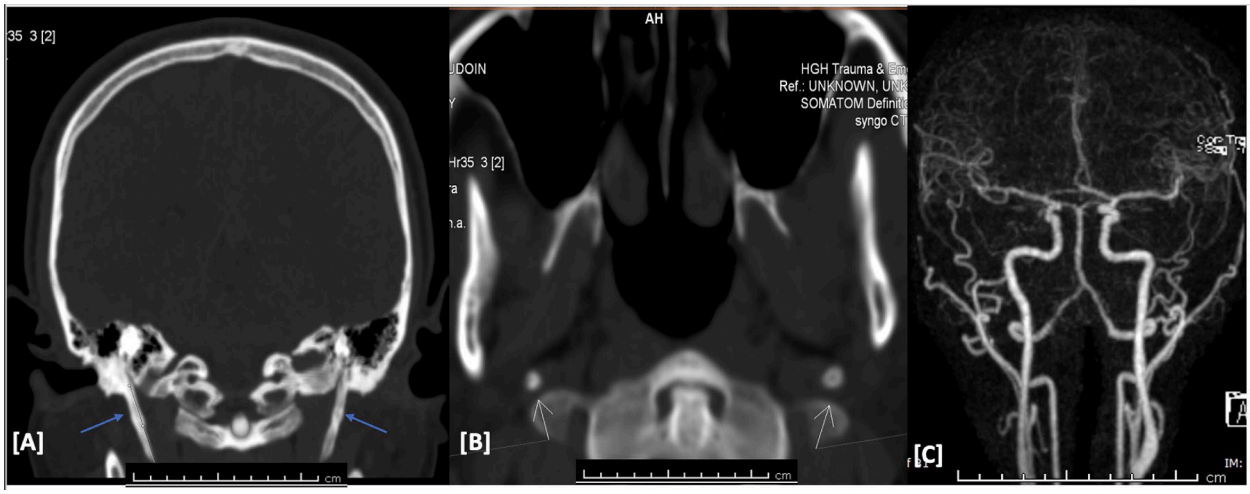

Fig. 3. [A–C] CT head- Case two bilateral elongated styloid processes. [A and B] Axial and Coronal reconstructed MIP CT images in bone window setting showing thickened elongated styloid processes bilaterally (Blue and white arrow). [C] CT angiogram showing normal vasculature with no evidence of stenosis or dissection.

development [3]. Revilla and Stuyt et al., proposed the process of endochondral ossification of Reichert's cartilage aids in developing the styloid process, stylohyoid ligament in utero by three month of fetal life which might explain variants in styloid process length and calcification. [3]. Perhaps Eagle's syndrome remains an overlooked and under-reported etiology of extracranial ICA dissection with most reported cases seem to highlight a pattern of vascular (Extracranial ICA) compromise triggered by neck manipulation [4]. In a recent literature review, extracranial ICA dissection was reported to constitute up to 39.1% of reported vascular Eagle's syndrome [4–6].

Reported clinical manifestations of Eagle's syndrome due to neurovascular structure compression can vary on a spectrum from cervicofacial neuralgia to vertigo, syncope, transient ischemic attack (TIA), and cerebral ischemia [4]. Reported variants of the classical subtype can manifest with otalgia, odynophagia, and recurrent paroxysms of headache which has been postulated to be due to the impingement of elongated styloid process or the ossified stylohyoid ligaments onto the traversing cranial nerve branches (Cranial Nerves V, VII, VIII, IX, X) [4,5]. Often certain maneuvers like neck manipulation and yawning can exacerbate it or even elicit cranial neuralgia (i.e: Glossopharyngeal neuralgia) [3–5].

The pathophysiology of vascular Eagle syndrome seems to correlate with a prolonged styloid process in length and its angulation as highlighted in a recent retrospective study on styloid process-related carotid dissection (SPRD) [6]. They demonstrated the relevance of the first cervical (C1) vertebral -styloid process distance to be shorted on ICA dissected side reinforcing its etiological implication [6]. Anatomically speaking, normal styloid process length is estimated to be 25–30 mm, with an increment in its length by 1 mm seems to increase the likelihood of developing ICA dissection by an odds ratio of 1.08, emphasizing it as a plausible differential in atraumatic CAD [6,7]. The mean age of vascular Eagle syndrome at onset is 49.2 years (range 38–80) year with no underlying vascular or connective tissue disorders, as observed in our two reported patients with Eagle's syndrome of variable presentations [7].

Evaluation of Eagle's syndrome is multifaceted in approach with emphasis on clinical symptoms, exclusion of local causes (temporomandibular joint disease, local pharyngeal masses or nearby compressing tumors), and radiological findings most accurately instigated via orthopantomogram (OPG) and three-dimensional computed tomography of the neck (3D CT) reconstruction and measurement of styloid process and ossified stylohyoid ligament [3]. In Vascular Eagle's syndrome vascular imaging with angiogram is warranted especially in cases of carotid dissection, venous or arterial stenosis/compromise as suspected in our patient (Case 1) for further assessment of ipsilateral side of styloid process elongation, angulation, and its relative relationship to ICA dissection [4]. Vascular Eagle's syndrome has been reported in literature across the spectrum of ages with cerebral ischemia in early adolescence and middle age that has been medically managed successfully in conjunction with angioplasty, stenting, and inevitably styloidectomy [5,9,10]. Our cases further empathize the relevance of prolonged styloid process in atraumatic ICA dissection and considering surgical management to prevent further recurrent dissections which can over-all improve outcomes in such strokes.

The consensus on the treatment of extracranial ICA dissection does not favor anti-platelet over anti-coagulation therapy, hence management is tailored for each patient [8]. Owing to under-reported cases of vascular Eagle's syndrome and the rarity of such entity, there is no general consensus on management [6]. However, styloidectomy (removal of elongated portion of styloid process) is considered the first-line treatment when medical management or endovascular intervention (vascular stenosis and thrombus retrieval) are inadequate to prevent further embolic phenomenon and vascular complication (i.e: dissection related pseudoaneurysms) [6,7]. There are two surgical approaches for styloidectomy: the intraoral approach (or transpharyngeal) and the extraoral approach, with the intraoral approach being preferred [11]. These surgical approaches take in consideration important landmarks and variant of trigeminal porus (TP) types near the Meckel's cave (ellipse and slit-like types) with regards the favorable surgical approaches [12]. Another consideration in the surgical approach nearby skull base and styloid process, are dural septation types pertaining to the jugular foreman and traversing cranial nerves like (glossopharyngeal nerve (cranial nerve [CN] IX), vagus nerve (CN X), and accessory nerve (CN XI)) [13]. Our two cases have been managed via a multidisciplinary approach with scheduled styloidectomy to alleviate cervical-facial neuralgia and ameliorate the risk of developing ICA dissection (case 2) or further cerebral compromise (case 1).

4. Conclusion

Eagle's Syndrome is a rare entity that is perhaps overlooked and labeled as a normal anatomical variant. However, it should be considered a possible etiology in atraumatic ICA dissection in patients with no discernable risk factors. Surgical intervention (styloidectomy) could avert risk of recurrent dissections and alleviate refractory pain and in patients with recurrent orofacial pain syndromes if other possibilities are excluded.

We believe more studies are warranted to further establish benefits and risks of styloidectomy. Hence, to conceivably establish guidelines to whom surgical intervention can possibly mitigate risk of ICA dissection and improve symptomatology and over all outcomes.

Ethics approval and consent to participate

This case report was approved by the Hamad Medical Corporation's Medical Research Center (MRC-04-23-195).

Consent for Publication

Written informed consent was obtained from the patient for publication of this case report, any accompanying images and photography. A copy of the written consent is available for review from the Editor of this journal.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on request.

Author contributions

All authors have read and approved the manuscript.
Writing the initial draft of the manuscript: AS, MAA, IA.
Conceptualization and supervision: YI, GM, AA.
Medical management of the case: MAA, AS, YI, GM, IA.
Critically revising the manuscript and literature review: AS, YI, AA.

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Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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