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Review Article

Eagle syndrome: An updated review

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ABSTRACT

Background: This work aims to review the current literature and our experience on vascular Eagle syndrome (ES) that can present misleading clinical presentations and better understand the possible therapeutic strategies.

Methods: We reviewed the existing literature on PubMed from January 1, 2017, to December 31, 2022, including the sequential keywords "*vascular* AND *Eagle syndrome*," "*vascular* AND *styloid syndrome*," "*vascular* AND *elongated styloid process*," "*vascular* AND *stylocarotid syndrome*," and "*Eagle syndrome* AND *carotid artery dissection*."

Results: 38 vascular ES cases, including our experience, were analyzed. The most frequent clinical onset was hemiparesis (*n* 21, 57%), but other regular clinical presentations were aphasia, loss of consciousness, amaurosis, headache, or a combination of the latter. Massive oral bleeding was reported only once in the literature before our case. Twelve patients were treated with only antiplatelet therapy, either single or double. Nine patients were treated with anticoagulation therapy only. In 14 patients, a carotid artery stent was used, associated with anticoagulation or antiplatelet therapy. In 17 cases, a styloid process (SP) resection was performed.

Conclusion: ES has many clinical presentations, and carotid artery dissection resulting in oral bleeding seems rare. Literature results and our experience make us believe that when dealing with vascular ES, the best treatment strategy is endovascular internal carotid artery stenting with antiplatelet therapy, followed by surgical removal of the elongated SP to prevent stent fracture.

Keywords: Eagle syndrome, Elongated styloid process, Internal carotid artery dissection, Stylocarotid syndrome, Vascular Eagle syndrome

INTRODUCTION

Eagle syndrome (ES) is a rare condition with different clinical presentations. First described by Eagle in 1937, it refers to various symptoms caused by the conflict of an elongated styloid process (ESP) – unilateral or bilateral – and/or calcified styloid ligament (CSL) with the surrounding anatomical structures. Originally, Eagle described this condition as a unilateral throat pain associated with the sensation of a foreign body in the throat.[11] Subsequently, the symptoms were classified into two main types depending on the involvement of either neural or vascular structures.[12] Odynophagia, facial and cervical pain, migraine, and dysphagia are usually caused

by the impingement of the lower nerves in the neck by the styloid process (SP). On the other hand, when talking about "vascular Eagle syndrome," authors refer to an ESP causing vascular compression, and often resulting in carotid artery dissection (CAD) and/or ischemia in the internal carotid artery (ICA) or middle cerebral artery (MCA) territories.[39,48] Many authors have described this syndrome, and the average length of the SP varies significantly in the literature. Conventionally, the SP is defined as elongated when it reaches 30 mm long.[4,26,32,34,40] The prevalence of ESP in the general population is around 4%, while symptoms are seen in only 4–10% of this group;[22] therefore, length itself is not always associated with symptoms. Other causes can contribute to the pathogenesis of this condition, such as soft-tissue disease, trauma, surgical insult (tonsillectomy), endocrine causes, or abuse of substances that can cause vasculitis-like damage facilitating compression and CAD.[44,50]

This review focused on the vascular form, a rare condition caused by the contact between ESP and/or CSL and the extracranial carotid artery. This conflict can lead to neurological symptoms (ischemia of downstream cerebral territories) due to compression, especially during rotational movement, or due to dissection and/or dissecting aneurysms of the carotid artery itself. We also present a case of a 48-year-old man without vascular risk factors contributing to mass oral bleeding and a later episode of transient dysarthria diagnosed with ES and treated with dual antiplatelet therapy (DAPT) therapy, endovascular flow-diverter (FD), and surgical SP removal. Since there is significant variability in clinical presentations and there is not a clear consensus on the management of this condition, we aim to summarize the current literature and to make a systematic review of cases of ES and CAD, considering articles from January 1, 2017, to December 31, 2022, expanding a previous study published in 2020 by Baldino *et al*.[5]

MATERIALS AND METHODS

We reviewed the existing literature on PubMed from January 1, 2017, to December 31, 2022, without restrictions about the paper publication status, according to the preferred reporting items for systematic reviews and meta-analyses (PRISMA) statement.[33]

We researched PubMed for the literature regarding patients with an ESP and/or ligament calcification causing either cervical ICA dissection or cervical ICA dissecting aneurysms. The following sequential keywords were used: "*vascular* AND *Eagle syndrome*," "*vascular* AND *styloid syndrome*," "*vascular* AND *elongated styloid process*," "*vascular* AND *stylocarotid syndrome*," and "*Eagle syndrome* AND *carotid artery dissection*."

Inclusion criteria included articles in English published between January 1, 2017, and December 31, 2022, and considering ES only associated with ICA dissection or dissecting aneurysms. Exclusion criteria were articles focused on ES presenting with clinical features other than ICA dissection, articles in different languages or only abstracts available, pure reviews (without new cases), or editorials.

We analyzed different features:

- Population characteristics such as sex and age
- Symptoms of onset
- Laterality and length of ESPs
- Initial and eventual additional treatment
- Time of follow-up.

RESULTS

Through literature research on PubMed, between January 1, 2017, and December 31, 2022, 51 articles were found using the words "*vascular* AND *Eagle syndrome*, 48 articles using the words "*vascular* AND *styloid syndrome*," 53 articles using the words "*vascular* AND *elongated styloid process*," ten articles using the words "*vascular* AND *stylocarotid syndrome*," and 31 articles using the words "*Eagle syndrome* AND *carotid artery dissection*." Excluding repetitions, 67 original articles were screened through analysis of title and abstract. Thirty-two articles were then, excluded based on inclusion criteria cited before. A total of 35 articles were analyzed through full-text reading, and four more articles were excluded due to non-interesting data. In summary, 31 articles were used for this review [PRISMA graph in Figure 1].

This paper reports an additional case of CAD associated with an ESP, treated at Belcolle Hospital in Viterbo in September 2018.

Our review of the literature shows that between 2017 and 2022, 37 cases of internal CAD have been caused by the vascular variant of ES. The demographic, clinical characteristics, treatment strategies, and follow-up of the patients are summarized in Table 1.[1,2,5,6,10,14-16,18-21,23,24,27,30,31,37,38,42-47,50-52,54,55,57]

The mean age of the patients was 48.2 years (range 30–72). In an article, one patient was described as a "teenager" but was not used for statistical evaluation. The prevalence was higher in men (n = 24, 69%) than in women (n = 11, 31%). In two cases, sex was not specified. In only one case, internal CAD was found occasionally, while an acute onset was described in all other cases. The most frequent clinical onset was hemiparesis (n = 21, 57%), but other frequent clinical presentations were aphasia, loss of consciousness, amaurosis, headache, or a combination of the latter. Bleeding from the oral cavity with hemoptysis was reported in only one case, resulting in a rare clinical presentation.

In the literature, there is a significant variability of treatment strategies. There is not a strong consensus on whether to prefer anticoagulation or antiplatelet therapy. In these cases, 12 patients were treated with only antiplatelet therapy,

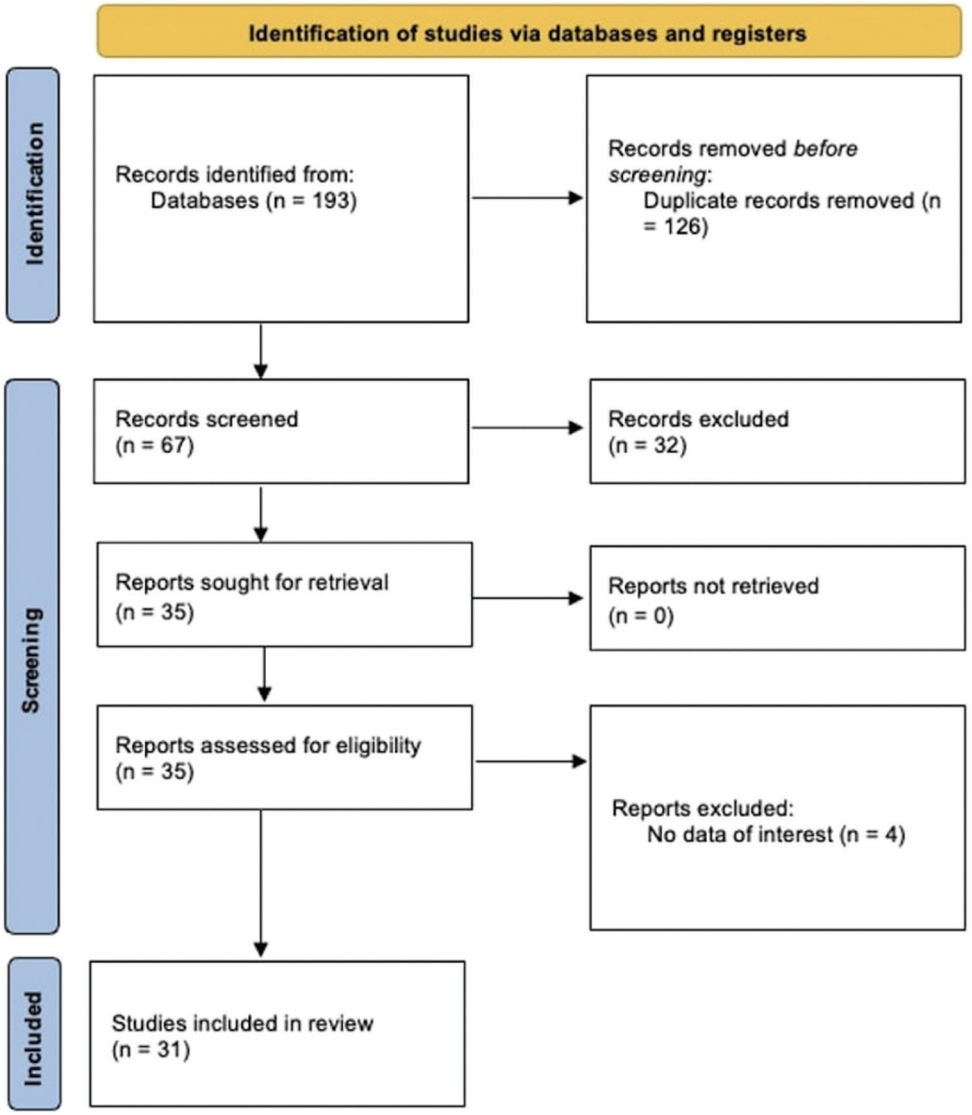

Figure 1: Preferred reporting items for systematic reviews and meta-analyses graph shows paper selection method.

either single or double. Nine patients were treated with anticoagulation therapy only, with previous thrombus aspiration in two of these cases. In 14 patients, a carotid artery stent was used, associated with anticoagulation or antiplatelet therapy. In two cases, thrombectomy of MCA occlusion was necessary before stenting. In three cases, angioplasty was used before stenting. In 17 cases, a SP resection was performed. Only in one case, SP resection was used as the only treatment, while, in all other cases, it was used as a secondary procedure after the resolution of the CAD to eliminate the mechanical cause and prevent recurrence.

Clinical case

In September 2018, a 48-year-old man without any history of vascular and neurovascular risk factors was admitted to our Emergency Room due to a sudden massive oral bleeding and a slight spatial and temporal disorientation. Contrast enhancement computed tomography (CT) scan showed a large portion of hypodense tissue with poor enhancement in the left retropharyngeal space, with an axial diameter of about 5 × 3 cm [Figure 2]. Such tissue medially dislocated the tonsils, imprinting the oropharyngeal aerial column, and involved the vascular space laterally, compressing the left internal jugular vein, but with regular size internal and external carotid arteries. Such hypodense tissue with mass effect is oriented toward different diagnoses, including head and neck neoplasm, infectious-inflammatory disorders, or bleeding carotid artery aneurysms. Video laryngoscopy examination highlighted an ab extrinsic imprint of the oropharynx on the left. A week after admission, the patient experienced an episode of transient dysarthria, and

Table 1: A comprehensive literature analysis of vascular Eagle's syndrome cases.

Articles	Year	Cases	Age	Sex	Symptoms	Laterality of ESPs	Stiloyd process lenght	Initial treatment	Additional tratment	Followup
Hebant *et al.*	2017	1	57	M	Aphasia, right facial central palsy	Bilateral	R: 33 mm; L: 31 mm	Single antiplatelet	None	2 months
Aydin *et al.*	2017	1	57	F	Left central facial palsy and left hemiparesis	Bilateral	-	Antiplatelet (not specified)	None	36 months
Subedi *et al.**	2017	1	47	F	Left upper extremity weakness	R	42 mm	Carotid artery stenting	SP resection scheduled	Not reported
Mann *et al.**	2017	1	39	F	Left hemiparesis	R	55 mm	Anticoagulation	Carotid artery stenting	5 years
Amorim *et al.*	2017	1	68	M	Aphasia, right inferior facial palsy	Bilateral	R: 38 mm; L: 46 mm	Single antiplatelet	Not reported	Not reported
Smoot *et al.*	2017	1	60	M	Left hemiparesis	Bilateral	R: 69 mm; L: 44 mm	Anticoagulation	Carotid artery stenting	Not reported
Jo *et al.*	2017	1	38	F	Headache and left perioral numbness	R	45 mm	Dual antiplatelet	SP resection	6 months
Zammit *et al.*	2018	1	45	M	Ipsilateral headache, facial sensory symptoms, right horner's syndrome	Bilateral	R: 40 mm; L: 42 mm	Dual antiplatelet	Not reported	Not reported
Tan *et al.*	2019	1	50	M	Right hemiparesis and aphasia	Bilateral	R: 32 mm; L: 31 mm	Trombectomy of accociated M1 trombus and dual antiplatelet	Stenting and angioplasty	6 months
Jelodar *et al.*	2018	1	40	M	Multiple episodes of ischemic strokes, slight right hemiparesis and dysphasia	Bilateral	-	Antiplatelet (not specified)	SP resection	8 months
Shimozato *et al.*	2018	2	48	M	Left hemiparesis	Bilateral	R: 33 mm; L: 37 mm	Carotid artery stenting and thrombectomy	Dual antiplatelet	21 months
			45	M	Aphasia and right hemiparesis	Bilateral	R: 30 mm; L: 31 mm	None	Carotid artery stenting and dual antiplatelet	4 months
Qureshi *et al.*	2019	1	42	F	Right hemiparesis and facial palsy and aphasia	Bilateral	R: 60 mm; L: 45 mm	Dual antiplatelet	None	3 months
Torikoshi *et al.*	2019	1	46	M	Left hemiparesis	Bilateral	> 40 mm bilat	Bilat carotid artery stenting	Bilat SP resection	3 years
Galletta *et al.*	2019	1	53	M	Left hemiparesis and facial palsy and dysarthria	Bilateral	35 mm bilat	Thrombus aspiration and LMWH	SP resection	3 months
Shindo *et al.*	2019	1	49	M	Left hemiparesis and dysarthia	R	31 mm	Anticoagulation and antiplatelet (not specified)	None	6 months
Michiels *et al.*†	2020	1	64	M	Right cervical swelling and hemoptysis, pain	Bilateral	R: 46 mm; L: 50 mm	SP resection	None	5 months

(Contd...)

Table 1: *(Continued).*

Articles	Year	Cases	Age	Sex	Symptoms	Laterality of ESPs	Stiloyd process lenght	Initial treatment	Additional tratment	Followup
Baldino *et al.**	2020	5	49	M	Left aphasia	Bilateral	-	Anticoagulation	Bilateral staged SP resection	1 year
			45	M	Left-sided amaurosis fugax	Bilateral	-	Anticoagulation	SP resection	5 years
			41	F	Occasional	R	-	Flow Diverting stent	Dual antiplatelet	4 years
			48	M	Right transient hemiparesis	L	-	Anticoagulation	SP resection	3 years
			56	M	Transient loss of conciousness	Bilateral	-	Anticoagulation	SP resection scheduled	6 months
Horio *et al.*	2020	1	46	F	Right hemiparesis and aphasia	Bilateral	R: 31 mm; L: 33 mm	Dual antiplatelet and left carotid artery stenting	SP resection	Not reported
Ikenouchi *et al.*‡	2020	1	30	M	Mild sensory disturbance in left limbs	Bilateral	R: 35 mm; L: 36 mm	Single antiplatelet	None	3 years
Kesav *et al.*	2020	1	49	M	Aphasia and right hemiparesis	Bilateral	R: 61 mm; L: 57 mm	Aspiration thrombectomy and LMWH	Dual antiplatelet	Not reported
Brassart *et al.*	2020	1	53	F	Left hemiplegia, tinnitus and palpitations	Bilateral	31 mm bilat	Anticoagulation	SP resection scheduled	1 year
Tanti *et al.*	2021	1	-	-	Loss of conciousness, left facial palsy, visual extintion and sensory loss	Bilateral	R: 74 mm; L: 70 mm	Dual antiplatelet	Patient refused stiloidectomy	3 years
Duarte-Celada *et al.*	2021	1	43	F	Headache and pharyngalgia, left Horner's syndrome	Bilateral	-	Angioplasty and carotid artery stenting and dual antiplatelet	Bilateral SP resection	2 years
Xhaxho *et al.*	2021	1	64	M	Loss of conciousness, headache, right eye amaurosis, right hemiparesis, neglect	Bilateral	-	10-days conservative therapy - not specified	SP resection	3 months
Selvadurai *et al.*	2022	1	teenage	M	Left hemiparesis and dysarthia	Bilateral	R: 34 mm; -	Thrombolisis and anticoagulation followed by antiplatelet therapy	SP resection and planned controlateral sp resection	4 weeks
Shi *et al.*	2022	1	30	M	Left hemiparesis, somnolence and urinary incontinence	Bilateral	R: 32 mm; L: 35 mm	Antiplatelet (not specified)	Patient refused stiloidectomy	Not reported
Lakner *et al.*	2022	1	38	-	Right hemiparesis and aphasia	Bilateral	R: 45 mm; L: 50 mm	Dual antiplatelet	SP resection scheduled	Not reported
Sasaki *et al.*	2022	1	37	M	Right amaurosis fugax	Bilateral	R: 32 mm; L: 34 mm	Dual antiplatelet	Embolization of associated aneurysm and carotid artery stenting	Not reported

(Contd...)

Table 1: *(Continued)*.

Articles	Year	Cases	Age	Sex	Symptoms	Laterality of ESPs	Stiloyd process lenght	Initial treatment	Additional tratment	Followup
Okada *et al.*	2022	1	72	M	Dysarthria and conciousness dysorder	L	32 mm	Anticoagulation and dual antiplatelet	Carotid artery stenting	Not reported
Matsukawa *et al.**	2022	1	38	M	Aphasia and right hemiplegia	L	33 mm	Trombectomy of associated MCA occlusion	Carotid artery stenting	6 months
Entezami *et al.*	2021	2	55	F	Left-eye vision loss	L	-	Dual antiplatelet	None	6 months
			46	F	Headache and right hand numbness, anisocoria, mild aphasia and dysarthria	L	-	Single antiplatelet	Angioplasty and ICA stenting - surgical resection of SP	6 months
Present case	2022	1	47	M	Sudden massive oral bleeding	Bilateral	R: 55 mm; L: 50 mm	Flow Diverting stent	Dual antiplatelet - surgical resection of SP	48 months

(*) cervical carotid artery dissecting aneurysm
(†) ICA blushing at angiography examination, without evidence of dissection
(‡) 3 y before left ICA dissection (ES not diagnosed during this episode). Patient with Ehlers-Danlos Syndrome. M: Male, F: Female, R: Right, L: Left, SP: Styloid process, ESPs: Elongated styloid processes, LMWH: Low molecular weight heparin

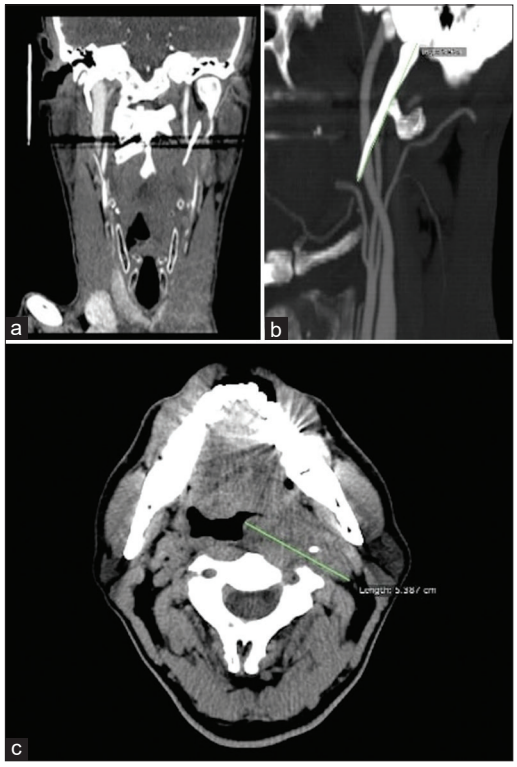

Figure 2: (a and b) Coronal and sagittal computed tomography (CT) scans show the elongated left styloid process causing internal carotid artery compression. (c) The axial CT scan showed hypodense tissue in the left retropharyngeal space with poor enhancement and diameters of about 5 × 3 cm.

magnetic resonance imaging of the head and neck region was performed. The examination better defined the tissue limits near the medium-distal extracranial tract of the left carotid artery, showing a possible aneurismatic dilatation of about 4.5 mm of the anterior wall of the artery. The patient underwent a CT angiography that confirmed the presence of a small bleb of about 3 mm in size, located at the top third of the left carotid artery at the apex of the left SP. The bone examination confirmed the presence of a longer SP bilaterally (about 50 mm on the left and 55 mm on the right) and a conflict on the left side, causing the compression of the homolateral ICA [Figure 3]. After multidisciplinary discussion, percutaneous endovascular treatment of the ICA dissection was performed, and an FD device (Streamline 5 mm × 25 mm) was placed [Figure 4]. DAPT was continued for six months. After 12 months, surgical removal of the left SP was obtained. The surgical procedure was performed in mono antiplatelet therapy. The patient fully recovered and remained asymptomatic, and at 48 months, the radiological examinations confirmed the presence of the previously implanted stent on the extracranial ICA, which appears normally positioned and patent [Figure 5].

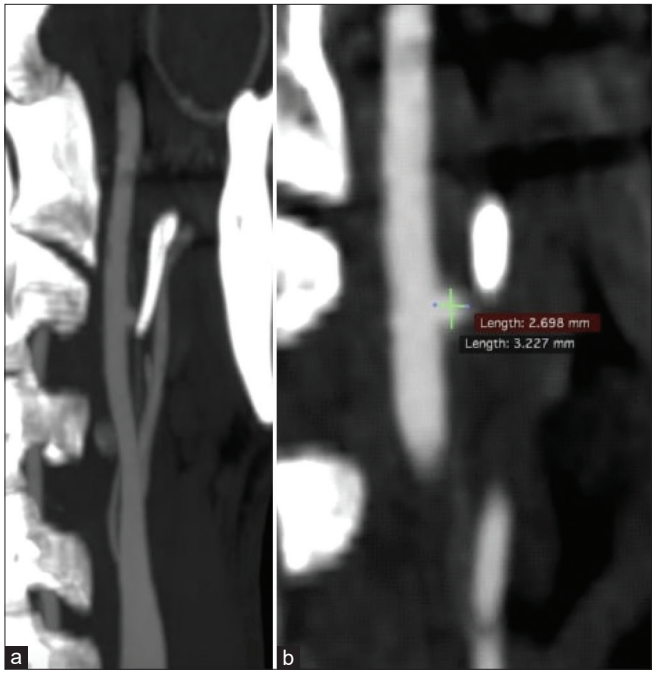

Figure 3: (a and b) A small bleb about 3 mm in size is shown, located at the top III left internal carotid artery at the height of the apex of the temporal style process.

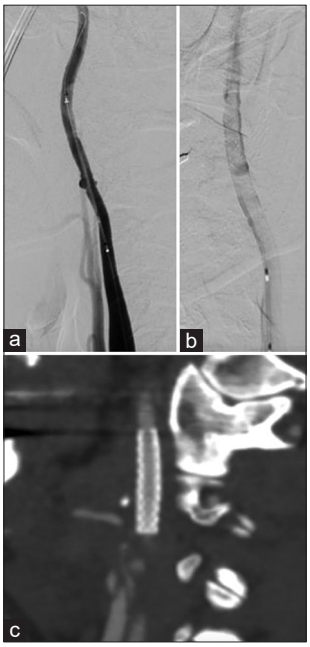

Figure 4: (a and b) Lateral angiography of the left carotid artery shows the placement of the flow-diverter device (Streamline 5 mm × 25 mm). (c) One month computed tomography scan shows stent placement with no complications.

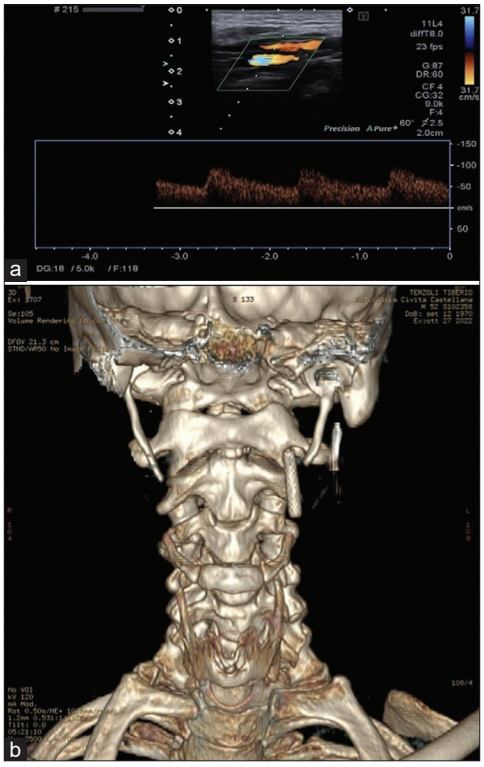

Figure 5: (a) The echo-color-Doppler examination of the left internal carotid artery documents the presence of the flow-diverter stent, which appears in the correct position and patent with an intra-stent sampled velocity index in range. (b) Postoperative 3D reconstruction computed tomography scan; reducing the styloid process length avoids impingement with the stent.

DISCUSSION

Massive oral bleeding is a rare and nonspecific clinical presentation related to traumatic events, upper gastrointestinal tract tumors or inflammatory processes, ulceration, rupture of esophageal varices, use of anticoagulation therapy, and many other underlying issues. When bleeding occurs in healthy young patients without comorbidities nor apparent triggers, it arouses concerns and attention. One rarely reported cause of oral cavity bleeding can be CAD associated with vascular ES.[25,31]

First described by Eagle in 1937, ES is a rare condition characterized by an abnormal length and/or slope of SP or a CSL.[11,12] The origin of an ESP or ossified styloid ligament may be identified in retained embryologic cartilage tissue from Reichert's cartilage, calcification of the stylomandibular ligament or expansion of osseous tissue at the origin of the stylomandibular ligament,[35] inflammation due to aging,[7] and reactive hyperplasia or metaplasia from trauma or simply as an anatomical variant without evidence or history

of trauma,[49] but the exact cause and etiopathogenesis remain unclear.

SP physiological length is around 20–25 mm, and 3 cm is usually the upper limit. A length of 4 cm is highly associated with symptoms, varying from pain in the lateral cervical area alone to severe vascular or nervous complications. In the presented case, SP length was 5 cm on the left side and 5.5 cm on the right. The incidence of elongated SP in the general population is 4–10%, but only approximately 4% of these cases are symptomatic; therefore, ES is <2% in the general population. As we can also notice in our case, even if the SP on the right side was longer, it did not conflict with neurovascular structures, highlighting that the length itself is insufficient to provoke the symptoms. In this small percentage of cases, the anatomical abnormality determines symptoms that define two pictures commonly recognized as "neurological variant" and "vascular variant" due to conflict with the adjacent anatomical structures. Indeed, in its course from the temporal bone posteriorly to the mastoid apex toward the maxillo-vertebro-pharyngeal recess, SP has a relationship with many important vascular (carotid arteries, and internal jugular vein) and nervous structures (facial, vagus, glossopharyngeal, and hypoglossal nerves). Based on clinical presentation, Eagle classified the syndrome into two subgroups: classic and vascular type. The classic type includes odynophagia, dysphagia, and/or cervical or facial pain elicited by head rotational movements. The vascular form, also known as stylocarotid syndrome, is a quite rare condition in which an elongated SP compresses the extracranial carotid artery and results in parietal or periorbital pain, and neurological symptoms such as headache, dizziness, transient visual loss, syncope, and stroke, often caused by compression or by the dissection of the carotid artery itself. In our case, oral bleeding was the first clinical presentation. This uncommon presentation has been described only once in the literature [31], and the exact mechanism remains unclear. Hemoptysis can probably be explained by blood leaks in the retropharyngeal recess and toward the surrounding cervical soft tissues.

Despite its benign nature, ES can lead to potentially severe complications. In the literature, many reports of transient ischemic attacks (TIAs) and strokes of the downstream territories of ICA are described, as an elongated SP has been recognized as a risk factor for CAD.[16,36,39,41,53]

The collection of medical, clinical, and radiological data can guide the diagnosis of vascular ES. Imaging, particularly a CT scan, is helpful in identifying the anatomical abnormalities and the presence of a neurovascular conflict. CT angiography is the gold standard and can provide further information regarding carotid flow, especially if stroke or dissection is suspected, and can be aided by 3D reconstructed images.[35]

Regarding treatment strategies, if patients with the classic form can be managed conservatively with analgesics, steroids, or local anesthetic, a surgical approach is preferred in cases of major vascular and neurological complications. A styloidectomy can be performed, and a transoral or transcervical method can be chosen.[3,9] Usually, the choice is dictated by the preference or experience of the surgeon. Since the association between CAD and elongated SP is uncommon, there are no exact guidelines regarding the best treatment.

Patients with symptomatic CAD presenting with ischemic stroke, TIA, retinal ischemia, or local symptoms only and without subarachnoid hemorrhage should be treated according to the ESO guidelines for managing extracranial and intracranial artery dissection.[8] When the patient presents with acute ischemic stroke and extracranial artery dissection, treatment with intravenous thrombolysis within 4.5 h from the symptom's onset should be performed. Mechanic thrombolysis is also recommended when a large vessel occlusion of the anterior circulation is encountered. Two multicentric randomized controlled studies[13,28] were analyzed regarding the use of anticoagulant versus antiplatelet therapy. The two groups had no statistical difference in the composite outcome of ischemic stroke, major bleeding, and death. The functional result was available in only one randomized controlled trial[13] and did not differ between the two groups. Subgroups of single versus double antiplatelet therapy did not show differences between the previous outcomes.

There is no firm recommendation on whether to use emergency stenting of the dissected carotid artery. One study[29] showed better successful reperfusion results with stenting than the non-stenting control group, but functional results did not differ.

We suggest that stenting of the ICA is advisable to achieve CAD correction in case of persistent ischemic symptoms, as in our case (dysarthria). Stenting can lower the risk of persisting vessel damage caused by the ESP while guaranteeing successful reperfusion of the downstream territories. As a result of recent literature, endovascular management of ICA dissection can provide good results, with a low rate of periprocedural complication and good long-term follow-up. [56] moreover, an association of surgical styloidectomy is recommended to eliminate the initial cause of neurovascular conflict and to prevent further damage to the implanted stent.[17]

CONCLUSION

ES has many clinical presentations, as evidenced by the multitude of nonspecific symptoms reported in the literature. The pathophysiologic mechanism of ES is related to the compression of the surrounding neurovascular structures.

The classic form of ES is described when nerve compression leads to craniofacial and cervical pain, and the sympathetic plexus's irritation around the ICA's cervical segment can cause vertigo and syncope. On the other hand, the vascular form is caused by direct compression of the extracranial ICA by the elongated SP, resulting in TIA or stroke and sometimes in CAD. CAD resulting in oral bleeding seems to be a rare clinical presentation, and the exact mechanism remains unclear. Literature results and our experience make us believe that when dealing with vascular ES, the best treatment strategy is endovascular stenting of ICA with antiplatelet therapy, followed by surgical removal of the elongated SP to prevent stent fracture.

We want to highlight that our experience may contribute to identifying a rare possible complication of vascular ES. Furthermore, the vascular form of ES should always be considered in the differential diagnosis in the presence of spontaneous nontraumatic CAD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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