The external carotid artery as a rare feeder of a spinal dural arteriovenous fistula causing cervical myelopathy: A review of the literature

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ARTICLE INFO

Keywords:
Ascending pharyngeal artery
Cervical myelopathy
External carotid artery
Occipital artery
Spinal dural arteriovenous fistula

ABSTRACT

Introduction: A causal relationship between SDAVF's and cervical myelopathy is exceedingly rare. 1–2% of these lesions are located at the cranio cervical junction of which 12% are caused by arterial feeders from the external carotid artery. A correct diagnosis can be challenging with a high rate of initial misdiagnosis.

Research question: Which aspects constitute the most important potential pitfalls in the diagnostic workup and treatment of SDAVF's with feeders from the external carotid artery causing cervical myelopathy?

Material and methods: We performed a PRISMA-guided review of the literature in which fourteen articles were included. We illustrate the diagnostic hazards through one of our own cases.

Results: SDAVF's at the cervical segment contain unique clinical and radiographic characteristics which differ from those elsewhere. Cervical myelopathy is caused by a SDAVF in 2.3% of cases. Pitfalls are numerous and diagnosis can be challenging, due to a broad differential diagnosis, potential isolated lower extremity involvement and absence of spinal cord edema on MRI. MR-alterations not always correlate with fistula localization.

Discussion and conclusion: A SDAVF should be part of the differential diagnosis in patients with subacute tetraparesis. When MRI shows signal alterations in combination with enlarged perimedullary vessels, a SDAVF should be suspected. Spinal angiography should include the vertebrobasilar system, as well as the internal and external carotid arteries. Early and adequate occlusion by means of an endovascular or neurosurgical approach of the draining radicular veins should be pursued. A multidisciplinary approach is key in the diagnostic workup and treatment of these patients.

1. Introduction

Spinal dural arteriovenous fistulae (SDAVF's) constitute a heterogeneous group of vascular malformations. Most lesions are acquired later in life and represent a pathological arteriovenous shunt between radiculodural, radiculomedullary arteries and perimedullary veins within the dura mater (Maimon et al., 2016; Onda et al., 2014). One to two percent of these lesions are located at the cranio cervical junction (Maimon et al., 2016; Onda et al., 2014; Hähnel et al., 1998; Hiramatsu et al., 2018; Ronald et al., 2020; Brinjikji et al., 2020; Geibprasert et al., 2009). Of these, only twelve percent of SDAVF's are believed to be caused by arterial feeders coming from the external carotid artery (Hiramatsu et al., 2018; Ronald et al., 2020; Brinjikji et al., 2020; Geibprasert et al., 2009).

Abbreviations: CASPR2, Contactin associated protein 2; CLIPPERS, Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids; CRP, C-reactive protein; DPPX, Dipeptidyl-peptidase-like protein; EMG, Electromyogram; GABAb, Gamma aminobutyric acid; LETM, Longitudinally extensive transverse myelitis; LGLI1, Glioma-associated oncogene 1; MEP, Motor-evoked potential; mGLUR1, Metabotropic glutamate receptor 1; MOG, Myelin oligodendrocyte glycoprotein; MRC, Medical research council; NMO, Neuromyelitis optica; PCR, Polymerase chain reaction; POEMS, Polyneuropathy organomegaly monoclonal gammopathy skin changes; PRISMA, Preferred reporting items for systematic reviews and meta-analyses; QUORUM, Quality of reporting of meta-analyses.

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https://doi.org/10.1016/j.bas.2021.100299

Received 17 August 2021; Received in revised form 30 August 2021; Accepted 21 September 2021

Available online 28 September 2021

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We performed a review of the literature, in which patients who developed a cervical myelopathy caused by a SDAVF were included. A total of 324 articles obtained from MEDLINE were identified. Search terms included “dural arteriovenous fistula” and “external carotid artery”. Additional studies were identified by checking reference lists (five articles). In total, fourteen articles were included. The entity is too rare to warrant a meta-analysis. This review of the literature is conform the 27-item checklist of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement.

2.2. Search strategy, eligibility criteria and data extraction

2.2.1. Search strategy

An adequate research question, based on the Patient-Intervention-Comparison-Outcome model, was formulated. Last search was performed on January 8, 2021. First author was responsible for database searches, abstract screening and quality assessment. Final inclusion was done after discussion with and approval by the senior author.

2.2.2. Information sources

We used the MEDLINE (PubMed) database. Reference lists were also checked for additional studies. Grey literature (research published in non-commercial form) was not examined due to arbitrary methodological reliability.

2.2.3. Eligibility criteria

Both retrospective and prospective studies were taken into account. Comments and technical notes were also included. Studies were screened on title and abstract, in which we focused on “external carotid artery”, “spinal dural arteriovenous fistula” and/or “cervical myelopathy”. Inaccessible or duplicate studies were excluded. Irretrievable information was left blank, meaning that authors were not contacted in case of incomplete or missing data. All studies were handled with a sufficient amount of strictness. All major forms of bias were checked when reading the individual included studies. No studies had to be excluded within the context of the predefined quality criteria.

3. Results

3.1. Study selection

Results are based on fifteen cases out of a total of 324 articles obtained from MEDLINE (fourteen cases extracted from previous literature and our case report). The sequence of consecutive steps used in our literature review is depicted by the QUORUM flow chart (Fig. 1). A total of fourteen articles met the final inclusion criteria. 310 articles were excluded for reasons provided in Fig. 1.

3.2. Case presentation

A 58-year old woman was referred to our hospital as a result of rapidly progressive tetraparesis. She presented to our hospital due to an acute brachial pain syndrome. In addition to a nonmendelian pain irradiation into her left arm, our patient experienced diffuse paresthesias in her left leg and mentioned a history of recent repetitive falls without loss of consciousness or urinary incontinence. She had never experienced this kind of symptoms previously nor did she mention any symptoms with respect to her right hemispheric.

There was no history of head or cervical trauma. Examination of our patient’s medical records revealed benign arterial hypertension, hypercholesterolemia, obesity, type 2 diabetes and recent weight loss (16 kg due to gastritis with negative gastric biopsies, PET-scan, unenhanced abdominal CT-scan and a gastroscopy showing a hyperemic gastritis). She had a blank neurological and neurosurgical history.

Despite a questionable left Babinski sign, her clinical examination at presentation was unremarkable. In particular, a normal sensory and motor examination, as well as normal discocaudal provocation tests were withheld. An unenhanced cerebral, cervical and lumbar CT-scan were performed, showing no abnormalities despite some coincidental degenerative changes. Therefore, in order to perform further investigations, she was hospitalized for intravenous pain medication, with a discocaudal pain syndrome as the initial preferential diagnosis.

The next day, an electromyographic (EMG) examination was performed, which showed normal MEPs as well as normal and symmetric cortical motor latencies. Despite a mild lateralization of the left tibial muscle, no EMG signs of pyramidal tract injury could be objectified. Clinical examination remained unaltered.

Two days after clinical presentation however, first clinical deterioration became visible, with a mild paresis of her left arm (MRG 4 in biceps brachii and deltoid muscles), bilateral impaired finger abduction and mild paresis in both legs. Bilateral positive Hoffman-Trömmer and Babinski signs were present. There were no changes in awareness nor were there any symptoms suspicious for cauda equina syndrome, despite mild urinary retention. Furthermore, our patient developed fever (38.1 °C).

A broad inflammatory examination was performed, with negative blood results (mildly elevated CRP of 13 mg/l [< 5 mg/l]), chest X-rays, an elevated white blood cell count and nitrite in a urinary sample (644/μl [< 25/μl]) and repetitive hemocultures. Urinary cultures eventually became positive for Pseudomonas and Enterococcus, both sensitive to Levofloxacin. Covid-19 screening was negative. Transthoral echocardiography showed no signs of endocarditis. An unenhanced CT-scan of her chest and abdomen showed no abnormalities, in particular no signs of sarcoidosis.

We performed a lumbar puncture, which showed an elevated white blood cell count (21/mm3 [< 5/mm3]), elevated glucose (72 mg/dl [40-70 mg/dl]), protein (274 mg/dl [15-45 mg/dl]) and albumin...
(2090 mg/l [< 350 mg/l]) with normal lactate (2.4 mmol/l [1.1–2.4 mmol/l]). Cultures, oligoclonal bands and Herpes/Varicella PCR were negative eventually. Spinal fluid cytology showed no malignant cells. Further blood tests were performed, showing normal copper (92 μg/dl [80–140 μg/dl]) and zinc (83 μg/dl [71–109 μg/dl]), as well as negative oncological/NMO/MOG/GABAβ/DPPX/CASPR2/mGLUR1/LGI1 antibodies.

An urgent MRI of her cervical spine was performed, showing a T2 and T2-fatsat hyperintense signal in the central aspect of the cervical spine with extension into the medulla oblongata and upper thoracic spine (Fig. 2). Capricious hypointens structures were seen on the anterior aspect of the edematous medulla oblongata and cervical spine. An EMG re-evaluation showed neurogenic contractions in the left biceps brachii, suspect for cervical myelopathy. We decided to transfer our patient to a monitored unit for further exploration. Since an infectious cause with fever was not excluded, we started with a combined intravenous therapy consisting of Amoxicilline, Ceftriaxone and Aciclovir. No steroids were given, since there is a potential worsening of clinical conditions in patients with SDAVF after steroids administration (Ma et al., 1976).

Since most likely we had to deal with a SDAVF, we performed an angiogram of the vertebrobasilar system (Fig. 3). No fistula could be seen, however. The same day, an MRI of the cervical spine with contrast was performed, showing mild contrast captation at the posterolateral aspect of the medulla oblongata and anterior aspect of the cervical spine.

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**Fig. 1.** The Quality of Reporting of Meta-analyses (QUORUM) flow chart illustrating the consecutive steps that were followed during our review of the literature. Reasons for exclusion are depicted in Table 1.

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**Fig. 2.** MRI without IV contrast at initial presentation. A: T2-weighted imaging in the sagittal plane with fast-spin echo (TSE). A hyperintense signal in the central aspect of the edematous cervical spine with extension into the medulla oblongata and upper thoracic spine can be seen. B: T1-weighted imaging in the sagittal plane with fast-spin echo (TSE). Dilated perimedullary veins at the anterior aspect of the medulla oblongata can be seen. C: T2-weighted imaging in the axonal plane with merged fast field echo (MFFE).
Fig. 4. MRI with IV contrast. A: T2-weighted imaging in the sagittal plane with fast-spin echo (TSE). B: T1-weighted imaging in the sagittal plane with fast-spin echo (TSE). Dilated perimedullary veins at the anterior aspect of the medulla oblongata can be seen. C: FLAIR-imaging in the axonal plane with turbo inversion recovery magnitude (TIRM). D: T2-weighted imaging in the axonal plane with fast-spin echo (TSE). E: T1-weighted imaging in the coronal plane with magnetization prepared rapid gradient echo imaging (MPRAGE). F: T1-weighted imaging in the axonal plane with magnetization prepared rapid gradient echo imaging (MPRAGE). Mild contrast captation at the posterolateral aspect of the medulla oblongata and anterior aspect of the cervical spine can be seen.

(Fig. 4). Multiple prominent vascular structures at the craniocervical junction were noted. However, due to a negative angiogram in combination with rapid clinical deterioration, inflammatory changes and fever, we also considered an inflammatory or infectious origin. Our differential diagnosis at that time consisted of a vascular malformation, tumor, transvers myelitis, NMO, LETM or CLIPPERS.

Unfortunately, clinical deterioration continued. Our patient developed a tetraparesis, with a pronounced left hemiparesis (elbow flexion/extension 2/3, wrist flexion/extension 2/2, shoulder abduction 1) and right hemiplegia. Moreover, she noticed a diffuse hypoesthesia in her entire body without a sensory level. Laboratory findings remained stable. The next day, there was a spontaneous clinical improvement, although the motor deficits in both arms and legs were still present.

Ten days after presentation, we performed a MRI of her brain and
fullspine. These showed no signs of demyelinisation nor signs of optic neuritis. A normal location of the conus medullaris and normal aspect of midthoracic and lumbar spine were seen. Eventually, we decided to perform another angiogram to inspect both internal and external carotid arteries (Fig. 5). This examination showed a fistula at the left border of the foramen magnum with arterial transosseus feeders coming from the left occipital, posterior auricular and ascending pharyngeal arteries with venous drainage via a dural vein in connection with the anterior and posterior spinal veins. Finally, the diagnosis of a SDAVF causing cervical myelopathy as the underlying cause of her symptoms was confirmed.

The next day, an embolization with Onyx 18 (18 cSt, which is a measure for kinematic viscosity) of the left external carotid and occipital arterial feeders was performed under general anesthesia. We thoroughly discussed the potential risks of using liquid embolization near the skull base with our patient. Postoperatively, we started with corticosteroids to reduce the risk of medullary edema, as well as heparinization to reduce the risk of medullary vein thrombosis. Unenhanced CT-graphic re-evaluation was reassuring. Furthermore, rehabilitation was arranged. One week after embolization, an MRI of the cervical spine was performed showing full regression of the dilated perimedullary veins and clear regression of edema at the medulla oblongata and cervical spine (Fig. 6). Progression of contrast captation was noted however, probably due to venous infarction.

Three months after the procedure, our patient is able to perform activities of daily living with both arms (brushing her teeth, combing her hair etc.) and standing up from a sitting position independently. Walking is still very difficult and she remains dependent on a walker for mobilization. A clinical evaluation after six months is scheduled.

3.3. SDAVFs and cervical myelopathy

Spinal dural arteriovenous fistulae (SDAVFs) constitute a heterogeneous group of vascular malformations. Most lesions are acquired later in life and represent a pathological arteriovenous shunt formed within the dura mater near the spinal root sleeve between radiculodural and radiculomedullary arteries on one hand and perimedullary veins on the other (Maimon et al., 2016; Onda et al., 2014). In contrast to AVMs, these lesions lack a nidus and are not contained within the spinal parenchyma (Alonso Fernández et al., 2008). They account for about 60–80% of all spinal vascular malformations with an incidence of 5–10 cases per million (10% of all intracranial vascular malformations) of which 1–2% are located at the craniocervical junction (Maimon et al., 2016; Onda et al., 2014; Hähnel et al., 1998; Hiramatsu et al., 2018; Ronald et al., 2020; Brinjikji et al., 2020; Gélberprasert et al., 2009). Moreover, most SDAVF are located midthoracic (60%) or at the upper lumbar spine (32%) [1 (Ronald et al., 2020; Kulwin et al., 2012)]. The upper cervical or craniocervical localization is more frequently encountered compared to the lower cervical spine (Gélberprasert et al., 2009). In the cervical region, these malformations are less frequently observed compared to other vascular malformations, such as intramedullary AVMs and epidural AVFs (Brinjikji et al., 2020). A causal relationship between SDAVF and cervical myelopathy is exceedingly rare (Maimon et al., 2016; Hähnel et al., 1998; Brinjikji et al., 2020; Kleeberg et al., 2010). In a large retrospective study, this was the case in about 2.3% of cases (6 out of 258...
3.4. Pathogenesis of SDAVF’s

The exact etiology remains unknown, although reopening of a thrombosed radicular vein or thrombophilia has been suggested (Maimon et al., 2016). In a large retrospective study, 28% and 9% of patients had previous spinal surgery (e.g. anterior cervical fusion) and spinal trauma, respectively. An association between SDAVF and surgery/trauma is not clear, however (Ronald et al., 2020). Myelopathy is probably caused by any or a combination of venous hypertension, edema or hypoxia due to a reduced arteriovenous pressure gradient (Hähnel et al., 1998; Alonso Fernández et al., 2008; Kulwin et al., 2012). An increase in spinal venous pressure leads to a reduced arteriovenous pressure gradient, causing a diminished normal drainage and venous congestion (Geibprasert et al., 2009). Furthermore, the shunt causes an arterialization of radicular veins, creating a reversal of blood flow into the perimedullary venous plexus, leading to venous engorgement, hypertension and ischemia (Maimon et al., 2016).

In general, SDAVF are more frequently encountered in middle aged and elderly men, as was the case in a large retrospective study of Ronald et al. as well as Brinjikji et al. and Terao et al., although pediatric cases have been reported seldomly (Maimon et al., 2016; Ronald et al., 2020; Brinjikji et al., 2020; Geibprasert et al., 2009; Alonso Fernández et al., 2008; Terao et al., 1976). In patients with associated cervical myelopathy, there was no significant difference in gender or age (Brunereau et al., 1996).

The arterial feeders draining into the spinal medullary veins most often originate from intracranial arteries, the vertebral artery, branches of the internal carotid artery (meningohypophyseal artery or tentorial branches), branches of the external carotid artery (ascending pharyngeal, posterior auricular or occipital artery) or intracranial DAVF (Borden and Cognard type V) (Hähnel et al., 1998; Kulwin et al., 2012; Terao et al., 1976). In patients with a cervical myelopathy, most fistulae are fed by an intracranial artery, where the venous sinuses are mostly identified as the site of the fistula (Hähnel et al., 1998). It is exceedingly rare that patients suffer from more than one SDAVF (0.5–4%) (Maimon et al., 2016; Ronald et al., 2020).

SDAVF’s at the cervical segment contain unique clinical and radiographic characteristics (Onda et al., 2014). Moreover, there is a substantial angioarchitectural heterogeneity between SDAVF’s and other vascular malformations (e.g. perimedullary AVF, epidural AVF, intramedullary AVM) located in the same region (Brinjikji et al., 2020). Many cervical SDAVF show a common medullary vein for multiple dural and intradural arterial feeders (Onda et al., 2014; Geibprasert et al., 2009). At last, shunt flow is believed to be higher in the cervical region, although the risk of hemorrhage is low (Onda et al., 2014).

3.5. Clinical presentation

Since initial symptoms can be nonspecific and insidious, a correct diagnosis can be challenging with a high rate of initial misdiagnosis (Maimon et al., 2016; Hähnel et al., 1998; Brinjikji et al., 2020; Kleeberg et al., 2010). In most cases, sensory disturbances form the first and predominant complaint at initial presentation (Maimon et al., 2016). In a large retrospective study, lower extremity paresis was the most common symptom (91%), followed by sensory disturbances (87%) (Ronald et al., 2020). Pain is present in 50–60% of patients (radicular pain was present in our patient) (Maimon et al., 2016; Ronald et al., 2020). If left untreated, patients are at risk of developing an irreversible necrotizing myelopathy, better known as Foix-Alajouanine syndrome (Terao et al., 1976). A cervical SDAVF can even present with lower extremity weakness without upper extremity involvement, as depicted in the case of Geibprasert et al., in which a cervical SDAVF gave rise to isolated venous congestion of the conus medullaris (Brinjikji et al., 2020; Geibprasert et al., 2009). Rarely, an intracranial subarachnoid hemorrhage can be observed (Alonso Fernández et al., 2008). Arterial feeders from the external carotid artery are associated with a lower risk of hemorrhagic presentation, however (Hiramatsu et al., 2018). In most patients, a combination of both upper and lower motor neuron signs is present.
Upper extremity involvement is rather rare. Bulbar signs caused by brainstem dysfunction, such as dysphagia, vertigo and orthostatic hypotension, are the last to appear, as was the case in our patient (Bru nerneau et al., 1996; Terao et al., 1976). Bulbar signs such as respiratory insufficiency and need for endotracheal intubation has been reported (Kulwin et al., 2012; Terao et al., 1976). Mild symptoms of myelopathy can be present even when MRI does not show any signs of spinal cord edema (Maimon et al., 2016). Most symptoms are dependent on the venous drainage pattern and venous impairment is directly related to spinal cord dysfunction, therefore classification is based on venous drainage (Brunerneau et al., 1996).

3.6. Diagnostic workup

MRI/MRA is most frequently the first examination performed when cervical myelopathy is suspected. However, most SDAVF do not lead to spinal parenchymal contrast enhancement and usually develops at a later stage due to damage of the blood-brain barrier (Maimon et al., 2016; Hähnel et al., 1998). Therefore, whenever MRI signal alterations in a combination with enlarged perimedullary vessels are seen, a SDAVF should be suspected (Hähnel et al., 1998). MRA has a sensitivity of 90% and has the advantage to localize the lesion, since it can depict the first veins that fill with contrast and can form a guide for later digital subtraction angiography (DSA) (Maimon et al., 2016). Experience with CT-angiography is too sparse to recommend this tool for diagnostic purposes, although it can be used for follow-up (Maimon et al., 2016).

DSA has a higher sensitivity compared to MRA in order to demonstrate the spinal arteries and their arterial feeders and should therefore be regarded as the gold standard in the detection of SDAVF (Maimon et al., 2016; Hähnel et al., 1998; Ronald et al., 2020). In about 6% of cases, no abnormalities are seen on routine MRI even though myelopathy is clinically present (Maimon et al., 2016). Therefore, it is recommended to perform a DSA when a high suspicion is present based on clinical grounds even when MRI findings are limited (Maimon et al., 2016; Ronald et al., 2020). Furthermore, it should be noted that fistula localization can not be made by MRI, since signal alterations and perimedullary vein dilatation are not always correlated with fistula localization, as depicted by Geibprasert et al. (Maimon et al., 2016; Geibprasert et al., 2009). It is recommended to include the external carotid arteries as well, since it may gave rise to arterial feeders, as was the case in our patient (Maimon et al., 2016). Since DSA does not have a 100% sensitivity (15% negative results in one study (Ronald et al., 2020)), in suspected cases in which a fistula is not identified, all runs should be re-read before a final conclusion is made (Maimon et al., 2016). Negative angiograms can be caused by fistulae with limited shunting due to venous hypertension, technical difficulty due to tortuous bloodvessels and fistulae remote from MRI-alterations (Ronald et al., 2020). In conclusion, to avoid incorrect or delayed treatment, DSA of the vertebrabasil and carotid systems should be performed (Maimon et al., 2016; Hähnel et al., 1998).

3.7. Treatment

Spontaneous SDAVF occlusion is very rare and should not be expected. Therefore, early and adequate occlusion must be pursued (Maimon et al., 2016). Treatment of asymptomatic patients remains controversial, although these patients should be followed closely, both clinically and radiographically (Maimon et al., 2016). The main goal of treatment is to occlude the draining radicular veins at the dural attachment by means of an endovascular (N-butyl-cyanoacrylate or Onyx) or neurosurgical approach (caterization or clipping) or a combined procedure (Maimon et al., 2016; Geibprasert et al., 2009; Alonso Fernández et al., 2008; Bernard et al., 2018). To avoid irreversible damage and Foix-Alajouanine syndrome, treatment should be instituted as quickly as possible (Terao et al., 1976).

The success rate of endovascular treatment is generally slightly lower compared to surgery (80% versus 98% according to some studies), although the former is mostly less invasive (Maimon et al., 2016; Ronald et al., 2020; Terao et al., 1976). In a large retrospective study, four out of 40 patients who underwent surgery needed retreatment (10%), while this was the case in one out of seven patients (14%) undergoing endovascular treatment (Ronald et al., 2020). This is in line with another retrospective study of Terao et al. (1976). Excellent results after surgery have been reported, even for cervical lesions (Kulwin et al., 2012). If applicable, surgery should be guided with electrophysiological monitoring, intraoperative angiography or indocyanine green video-angiography (Onda et al., 2014). Due to a lack of randomized controlled trials, retrospective nature and small study populations, comparison of both treatment options should be performed with caution and all cases should be discussed at a multidisciplinary team meeting.

3.8. Prognosis

Prognosis depends on both severity and duration of the disease, although these associations are arbitrary according to some authors (Maimon et al., 2016; Ronald et al., 2020). In a large retrospective study, only symptom duration before treatment and history of initial misdiagnosis were significantly associated with a worse prognosis (Ronald et al., 2020). Half of untreated patients will become disabled within three years of diagnosis (Ronald et al., 2020). In general, 80% of patients show clinical improvement after treatment, while 4.5–11% show some form of clinical deterioration despite treatment (Maimon et al., 2016). It is still very difficult to prognosticate whether or not a patient will respond to treatment (Ronald et al., 2020). Generally, motor symptoms show greater improvement compared to sensory deficits, with a significant improvement in ALS gait scores in most patients (Maimon et al., 2016; Ronald et al., 2020; Terao et al., 1976). 35% and 54% of patients report improvement or stabilization of urinary function, respectively (Ronald et al., 2020). Follow-up based on clinical grounds and MR-imaging is recommended (Maimon et al., 2016). Persistent signal abnormalities or swelling of the spinal cord, even after complete occlusion of arterial feeders, is not uncommon (Hähnel et al., 1998; Terao et al., 1976). When no clinical improvement is seen or MRI does not show improvement, urgent angiography should be performed to rule out reopening of the fistula, neoangiogenesis of the collateral circulation or to screen for an infrequent second fistula, which has been reported by Terao et al. (Maimon et al., 2016; Terao et al., 1976).

4. Discussion

Since cervical myelopathy is more likely to be caused by trauma, neoplasm or myelitis, making a correct diagnosis can be challenging, as was the case in our patient (Hähnel et al., 1998; Ronald et al., 2020; Bernard et al., 2018). The rate of initial misdiagnosis is high, ranging from 35 to 81% (Maimon et al., 2016; Ronald et al., 2020). Other possible causes include Guillain-Barré syndrome, multiple sclerosis, neuro-myelitis optica and POEMS syndrome (Ronald et al., 2020).

In a large retrospective study, only 12% of SDAVF at the cranio-cervical junction were caused by an arterial feeder from the external carotid artery (Hiramatsu et al., 2018). In our patient, the ascending pharyngeal, posterior auricular and occipital arteries constituted the arterial feeders with a transosseus connection with the stylomastoid artery. All of these are branches from the external carotid artery. Normally, the ascending pharyngeal artery supplies the pharyngeal constrictor muscles, while the posterior auricular and occipital arteries supply the scalp posterior to the auricle. The stylomastoid artery is a branch of the posterior auricular artery which enters the skull through the stylomastoid foramen and is responsible for perfusion of the tympanic cavity and semicircular canals.

When cervical myelopathy is suspected, MRI/MRA is most frequently the first examination performed. In contrast to Hähnel et al., which reported that an extension of signal abnormalities into the medulla...
oblongata is more likely due to an intracranial fistula, this was not the case in our patient (Hahnel et al., 1998).

Depending on local practice and localization of the fistula, most authors agree that surgery should only be considered in selected cases in which the fistula is located posteriorly and easily accessible (Alonso Fernandez et al., 2008). Since the fistula was located anteriorly, we decided to perform embolization. In our hospital, most patients with a posteriorly located thoracic or lumbar SDAVF will be treated by means of a surgical approach. The usage of liquid embolization near the skull base with risk of causing additional spinal ischemia due to embolization of small perforating arteries was thoroughly discussed with our patient. Moreover, we considered the risk of performing additional damage to the tympanic vasa nervorum when accidently embolising the stylomastoid artery.

4.2. Limitations

Time to follow-up of our patient is rather limited, although this was not the main focus of our review.

5. Conclusion

Although rare, a SDAVF should be part of the differential diagnosis in patients with subacute tetraparesis, especially when clinical course is fluctuating. In particular, when MRI shows signal alterations in combination with enlarged perimedullary vessels, a SDAVF should be suspected. Classic spinal angiography constitutes the vertebrobasilar system and all spinal arteries to detect the fistula, in seldom variant cases originating from the external carotid artery with collaterals to the spinal cord. Therefore, DSA should always include both internal carotid, external carotid and vertebrobasilar arteries. This examination should be performed when a high suspicion is present based on clinical grounds even when MRI findings are limited. In suspected cases in which a fistula is not identified, all runs should be re-read before a final conclusion is made. Early and adequate occlusion by means of an endovascular or neurosurgical approach of the draining radicular veins at the dural attachment should be pursued. When no clinical improvement is seen after the procedure, urgent angiography should be performed to rule out reopening of the fistula. Finally, a multidisciplinary approach is key in the diagnostic workup and treatment of patients with a suspected SDAVF.

Financial disclosure
None.

Transparency statement
The manuscript is an honest, accurate and transparent account of the study being reported. No important aspects of the study have been omitted. PRISMA guidelines and CARE guidelines were followed.

Patient and public involvement statement
There was no patient and public involvement.

Submission declaration
We declare that this work has not been published previously nor under consideration for publication elsewhere. If accepted, it will not be published elsewhere in the same form or any other language without the written consent of the copyright-holder.

CRediT authorship contribution statement

Senne Broekx: Conceptualization, Formal analysis, Methodology, Validation, Visualization, Writing – original draft, Writing – review & editing.
Rik Houben: Conceptualization, Validation.
Luc Stockx: Data curation, Resources, Validation.
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Geert Gelin: Data curation, Investigation, Resources, Validation.
Frank Weyns: Data curation, Investigation, Resources, Validation.
Tom De Beule: Conceptualization, Data curation, Investigation, Methodology, Resources, Validation, Supervision.

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