Dural Arteriovenous Fistula and Foix-Alajouanine Syndrome: Assessment of Functional Scores with Review of Pathogenesis

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OBJECTIVE: **To assess the use of functional scores in the evaluation of patients with dural arteriovenous fistula and Foix-Alajouanine syndrome.**

METHODS: **We systematically surveyed the literature to identify relevant patients. Aminoff-Logue Scale (ALS) and modified Rankin Scale (mRS) scores were ascertained and combined to form a novel functional score, the Aminoff-Rankin Composite (ARC) score. We compared functional scores between surgery and embolization groups and ran one-sided point-biserial analyses to test our expectation that positive correlations exist between functional scores and treatment outcomes. Finally, we reviewed the pathogenesis of dural arteriovenous fistula formation.**

RESULTS: **The quantitative synthesis included 18 patients. Surgery alone was performed in 11 patients (61.11%); 7 patients underwent embolization alone (38.89%). There were no significant differences in functional scores or symptom outcomes when we compared surgery to embolization. The pre-intervention ALS gait, mRS, and ARC scores were correlated with improved symptoms ($r_{pb} = 0.43$, $P = 0.04$; $r_{pb} = 0.47$, $P = 0.02$; $r_{pb} = 0.48$, $P = 0.04$, respectively). In patients whose symptoms were improved, post-intervention ALS gait and micturition scores (2.55 vs. 4.43, $P = 0.02$ and 1.09 vs. 2.71, $P = 0.01$, respectively) and post-intervention ARC scores (6.66 vs. 11.57, $P = 0.01$) were on average lower than in patients whose symptoms were unimproved.**

CONCLUSIONS: **We believe that patients with dAVF and Foix-Alajouanine syndrome present with worse function (higher functional scores) as a result of an acute myelopathic episode, and that if diagnosed and treated appropriately, will experience some level of symptom improvement that is evidenced by reduced post-intervention functional scores.**

INTRODUCTION

Dural arteriovenous fistula (dAVF) can manifest as an insidious spastic-to-flaccid paraplegia, ascending sensory level, and loss of sphincter control with paroxysmal exacerbations. This classic presentation is known as Foix-Alajouanine syndrome. In 1926, Foix and Alajouanine published their French treatise *La myélite nécrotique subaigue*, which chronicled the clinical course of 2 young men with progressive, fatal myelopathies of unknown etiologies.[1] Pathologic analyses of the spinal cords revealed enlarged coronal venous plexuses and marked hypertrophy of the dorsal intradural veins. The impressions were endomesovasculitides of unknown origins.[2] Foix and Alajouanine[1] are credited with the first report of what would come to be known as Foix-Alajouanine syndrome, and the etiology (dAVF) was described many decades later.[3]

Foix-Alajouanine syndrome is well-recognized among neurosurgeons. However, there are some questions that remain unanswered. First, what is the optimal therapeutic strategy for dAVF? Second, what is the utility of functional scores in the clinical evaluation of patients presenting with Foix-Alajouanine syndrome? To address these questions, we collected and analyzed disaggregated data from published studies that described patients with dAVF and Foix-Alajouanine syndrome. Aminoff-Logue Scale (ALS) and modified Rankin Scale (mRS) scores were ascertained and combined to form a novel functional

Key words
- Arteriovenous fistula
- Arteriovenous malformations
- Spinal cord disease
- Venous congestion

Abbreviations and Acronyms
ALS: Aminoff-Logue Scale
ARC: Aminoff-Rankin Composite
AVM: Arteriovenous fistula
AVF: Arteriovenous malformation
dAVF: Dural arteriovenous fistula
mRS: Modified Rankin Scale

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score, the Aminoff-Rankin Composite (ARC) score. We compared surgery and embolization groups and tested our expectation that a positive correlation exists between functional scores and treatment outcomes. Finally, we reviewed dAVF pathogenesis.

MATERIALS AND METHODS

Preferred Reporting Items for Systematic Reviews and Meta-Analyses Protocols[4] were adhered to throughout this study. This systematic review is registered in the PROSPERO international prospective register of systematic reviews of the National Institute for Health Research. Six popular databases were searched through January 2017 using 10 strategic search term combinations (Table 1). Two of the authors (C.L. and L.K.C.) independently performed each stage of the search. English, full-text clinical studies and trials, case reports, and reviews were included.

Data Collection and Analysis

Adults (>17 years old) with progressive myelopathy and a diagnosis of spinal dAVF or AVM were included. Patient and clinical characteristics were disaggregated from the identified studies. Management of the fistula involved surgical excision or embolization. Patients who underwent a combined approach were excluded. ALS,[5] mRS,[6] and ARC scores were ascertained. Symptoms were dichotomized as improved or unimproved based on reduced functional scores after surgery/embolization or explicit report of symptom outcomes. We compared functional scores between surgery and embolization groups and ran one-sided point-biserial analyses to test our expectation that positive correlations exist between functional scores and treatment outcomes. All analyses were performed using SAS Studio (SAS Institute, Cary, North Carolina, USA) and tested at an α of 0.05.

RESULTS

We screened 714 articles. Nine studies that described 18 patients were included. The average age of patients was 52.78 years ± 13.17. The most common motor findings were weakness (44.44%) and paraplegia (33.33%). The most common sensory findings were hypoesthesia (55.56%) and saddle anesthesia (22.22%). The most common autonomic findings were incontinence (40.00%) and retention (40.00%). The most common reflex findings were areflexia (37.50%) and hyperreflexia (37.50%). The average duration of symptoms was 22.46 months ± 49.35.

Surgery alone was performed in 11 patients (61.11%), and embolization alone was performed in 7 patients (38.89%). The average follow-up duration was 52.67 months ± 71.15. There were no significant differences in functional scores or symptom outcomes when we compared surgery to embolization. The average pre-intervention ALS gait and micturition scores were 3.56 ± 1.89 and 2.33 ± 1.14, respectively. The average post-intervention ALS gait and micturition scores were 3.28 ± 1.96 and 1.72 ± 1.23, respectively. The average pre-intervention and post-intervention mRS scores were 3.77 ± 1.11 and 3.50 ± 1.58, respectively. The average pre-intervention and post-intervention ARC scores were 9.67 ± 3.61 and 8.50 ± 4.30, respectively.

The pre-intervention ALS gait score was correlated with improved symptoms ($r_{pb} = 0.43$, $P = 0.04$). The pre-intervention ALS micturition score was not correlated with improved symptoms ($r_{pb} = 0.34$, $P = 0.08$). The preintervention mRS score was correlated with improved symptoms ($r_{pb} = 0.47$, $P = 0.02$). The preintervention ARC score was correlated with improved symptoms ($r_{pb} = 0.48$, $P = 0.02$). In patients whose symptoms were improved, post-intervention ALS gait and micturition scores (2.55 vs. 4.43, $P = 0.02$ and 1.09 vs. 2.71, $P = 0.01$, respectively) and post-intervention ARC scores (6.66 vs. 11.57, $P = 0.01$) were on average lower than in patients whose symptoms were unimproved.

Table 1. Search Term Combinations

"Foix-Alajouanine"
"Foix-Alajouanine syndrome"
"venous" AND "congestive" AND "myelopathy"
"myelopathy" AND "dural" AND "fistula" AND "AVF"
"myelopathy" AND "venous" AND "malformation" AND "AVM"
"spinal" AND "spontaneous" AND "thrombosis" AND "fistula"
"spinal" AND "spontaneous" AND "thrombosis" AND "malformation"
"subacute" AND "necrotic" AND "myelopathy"
"subacute" AND "necrotic" AND "myelitis"
"angiodysgenetic" AND "necrotizing" AND "myelopathy"

AVF, arteriovenous fistula; AVM, arteriovenous malformation.

DISCUSSION

Pathophysiology

Although the origin of dAVFs is unknown, the lesions are thought to be acquired.[6-10] This theory is based on patient age, male predilection, absence of associated vascular anomalies, and the development of postoperative or traumatic intradural fistulas.[9-12] Prieto et al.[7] acknowledged that microtears of radiculomeningeal arteries sustained during collisions may favor fistula formation in the highly mobile cervical spine, but this conflicts with the observation that dAVFs are most commonly located in the thoracic region.[13-16]

The creation of a fistula initiates a cascade of events that alters the angioarchitecture and hemodynamic parameters of the intrathecal venous system. The fistula is typically fed by a dural branch of the intervertebral artery. Blood is diverted through the fistula under high pressure into the arterialized medullary vein, and this results in conformational changes to elastic and collagen fibers of the vascular wall. In principle, these changes are thought to occur along 3 axes: axial length, circumference, and radius.[17] Macroscopically, the changes are translated into elongation, dilation, and tortuosity of the vessels of the coronal venous plexus (Figure 1).[6,18]

The fistula is drained by a single, valveless medullary vein. The vein traverses the subarachnoid space, penetrates the dura mater adjacent to the nerve root, and then empties into the epidural venous plexus.[19] The dural penetration serves as a functional