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A Case of Synchronous and Metachronous Multiple Spinal Arteriovenous Fistulas Coexisting with Lumbar Lipomyelomeningocele: Literature Review

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Spinal arteriovenous fistula; Lipomyelomeningocele; Metachronous; Synchronous

1. Abstract

1.1. Objective: The concurrence of spinal dural Arteriovenous Fistula (AVF) with spinal dysraphism is extremely rare. This coexistence is suggestive of a causal relationship and should encourage science to study their common pathogenesis. We present a unique case in which a lipomyelomeningocele and multiple synchronous and metachronous AVFs occurred together; contributing to discuss about pathophysiological theories.

1.2. Methods: The authors retrospectively analyzed cases in literature involving patients with this dual coexistence. We also revised the documented cases of multiple synchronous and metachronous AVFs. Different pathophysiological theories have been analyzed and gathered.

1.3. Results: The research revealed 20 patients with AVF and dysraphism; Only 4 were described as multiple and among them the totality were reported as synchronous fistulas. On the other hand, 19 and 11 cases of multiple synchronous and metachronous AVFs have been reported respectively. According to recent theories, this coincidence seems a mixed result of incomplete regression of mesenchymal tissue during the embryological development and the evolutive spinal cord tethering caused by the lipoma which would impair venous drainage and create an unbeatable atmosphere for fistulas genesis. In addition and referring to the recent theory about multiple AVFs; the existence of one single AVF can promote the formation of multiple AVFs during time.

1.4. Conclusions: It is conceivable that there is a pathophysiological link between dysraphism and AVFs. The understanding of their association will lead the physicians to take the best decision in terms of diagnostic and therapeutic management. Delayed neurologic deterioration in a patient with dysraphism should strongly suggest the possibility of such an association.

2. Introduction

Spinal dural Arteriovenous Fistula (AVF) represent 80 % of all spinal vascular malformations while dysraphic abnormalities of the neuroaxis are also the most frequent congenital malformations of the central nervous system [1, 10, 12]. The AVF is considered to be an acquired lesion with natural history of progressive onset of neurological deficit, rarely present in individuals younger than 40 years.

The occurrence of multiple AVFs is rare. Synchronous AVFs represent those lesions that are happening simultaneously in different locations. True metachronous AVFs, defined as those different located lesions that are diagnosed in a temporal gap after the first AVF treatment and initial short period clinical improvement, are even more uncommon. [1, 2].

Neural Tube Defects (NTDs) represent a wide spectrum group of brain and spinal cord malformations, divided in two categories: open and closed spina bifida. In the case of closed spina bifida

its spectrum ranges from a simple skin stigma to the presence of Lipomyelomeningocele (LPM).

In this study we report, for the first time in our knowledge, a unique case of multiple synchronous and true metachronous AVFs in a single patient with lumbar dysraphism. Furthermore, we contribute to speculate on the possible related pathophysiologic origin of this exceptional association and discuss its diagnostic and therapeutic management.

3. Case Report

A 37-year-old man with a history of an asymptomatic lumbar L2-L4 dorsal LPM referred a 1-year period of progressive bilateral leg weakness with gait impairment, increasing drug-resistant back pain and sexual and bladder dysfunction. Neurological examination demonstrated mild paraparesis (4/5) with bilateral patellar hyperreflexia and positive Babinski sign.

Magnetic Resonance Imaging (MRI) showed L3 to L5 dysraphism related to L2-L4 intradural dorsal lipoma attached to the conus medullaris with extradural and extravertebral extension in association with tethered cord (L3) and significant spinal cord edema ascending up to D12. T2-weigthed MRI didn´t show the presence of direct suspicious signs of AVF.

Given the suspicion of vascular malformation, the patient underwent a selective spinal angiography disclosing the presence of multiple and synchronous AVFs at right L4 and left L3 levels, that were respectively supplied by bilateral lumbar segmental branches coming from the common iliac arteries. The selected treatment was endovascular embolization using Onyx injection, obtaining complete occlusion of both afferent branches (Figure 1).

During the initial follow-up, the patient reported progressive neurological improvement, regaining independence for his daily activities. Spinal MRI performed 1 month later revealed a clear improvement of previous myelopathy. Four months later he presented mild worsening of his gait and bladder impairments. Control MRI revealed increased myelopathy. A second spinal angiography with Glubran mixed with Lipiodol showed a new left L2 metachronous AVF. Incomplete endovascular occlusion was achieved due to small vessels caliber (Figure 2).

Surgical treatment with LPM resection and AVF disconnection was then indicated. Under general anesthesia and intraoperative neurophysiological monitoring L2L5 laminectomy was performed; LPM was circumferentially dissected and intradural exploration revealed tight attachment between the conus medullaris and the lipoma, which was grossly resected, followed by coagulation and disconnection of the AVF.

During follow-up, the patient's motor and impaired bladder symptoms improved considerably while sensory symptoms remained unchanged. Spinal MRI and angiogram performed at 6 months showed significant improvement in spinal cord edema and no evidence of residual AVF. Nowadays the patient is able to walk without assistance and has punctual urinary urgency episodes.

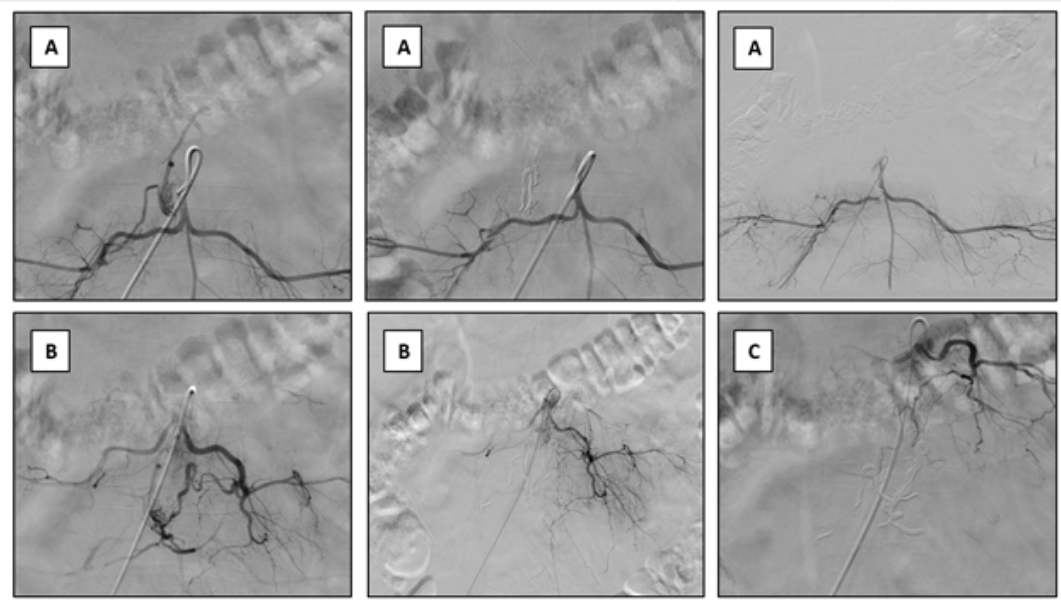

Figure 1: A, Embolization with Onyx of rigth L4 afference. There is an exclusión of the contribution even if good penetration is not achieved. B, After L4 afferential embolization, there is a recruitment of the fistula due to afferents coming for left L3. In these angiograms Left L3 fistula can be seen before and after the embolization with Onyx. C, At the end of the procedure there are no afferences coming from left L2 branch.

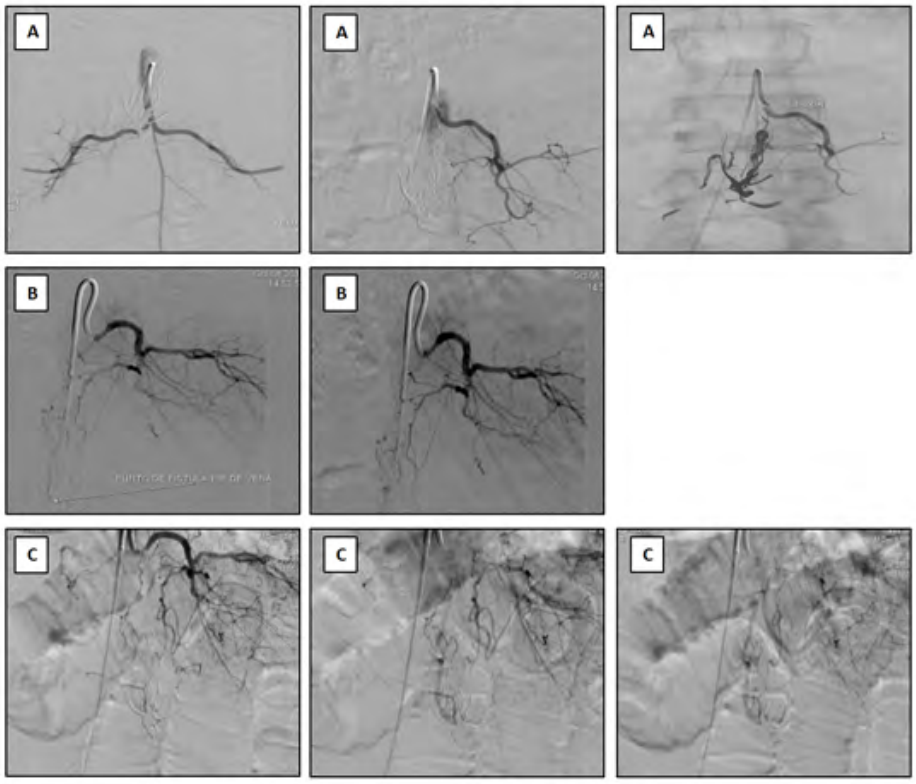

Figure 2: A, Segmental control is carried out to study previously embolized right L4 and left L3 fistulas without identifying contributions to the fistula. B and C, The study shows new recruitment of the fistula by non-catheterizable small afferent branches coming from left L2 fistula

4. Discussion

Spinal lipoma is a congenital disorder within the spina bifida spectrum which develops from a disturbance in the separation of the neuroectoderm from the cutaneous ectoderm and subsequent fusion with adipose tissue coming from mesoderm origin. These lipomas are normally diagnosed at birth and the symptoms are not usually developed until early adulthood due to progressive spinal cord tethering [4].

AVF is believed to be an acquired disorder which tends to present approximately in the sixth decade of life [7]. Several reports have described AVF secondary either to trauma or surgery [12-15]. Previous studies suggest that their formation might be related to venous outflow obstruction in the perimedullary veins due to thrombosis that leads to venous hypertension and spinal cord ischemia; promoting the right environment for the production of angiogenic factors and the development of AVFs [4, 7].

The association between AVF and spinal dysraphism at the same level is extremely rare. Only twenty similar cases have been described in the literature and all of them happened in adulthood (Table 1). Some authors argue that this coincidence might be the result of incomplete regression of mesenchymal tissue during a premature disjunction of the neuroectoderm which would lead to the migration of the periaxial mesoderm into the developing neural tube [4, 11]. While most of the pluripotent mesoderm would originate fat, they may also produce other tissues including blood vessels promoting abnormal angiogenesis in the lipomatous tissue proximal to the dural defect [6, 10]. As a final consideration to this physio-pathological theory, it is speculated that the lipoma and its evolutive spinal cord tethering would impair venous drainage, elevating the venous hypertension and clinical presence and development of AVF [6, 7, 11, 12]. Therefore, the type of AVF related to the spinal dysraphism would be an acquired entity supported, based and facilitated by a congenital disorder [4, 17]. In other words, its etiology may also be considered as a shared embryological origin with spinal dysraphism and should therefore be evaluated in our opinion as a different subtype of AVF [4].

Referring to clinical presentation, when these two disorders occur at the same time clinical overlap appears to be evident. Even if symptoms can be worsened with physical efforts and intra-abdominal pressure increasing activities in the case of AVF [5, 7, 12]; progressive onset of lower back pain, sensorimotor impairments and bladder disturbances are not reliable data to make difference between both entities. In this type of adult patients with this rare double coincidence, AVF should be considered as the potential and most probable cause of increasing symptoms [7, 13].

Talenti et al mentioned the importance of taking special attention to T2-weigthed MRI images where the presence of tortuous flow-voids on the spinal cord surface and new documented progressive myelopathy can suggest indirectly the presence of AVF [5, 13]. Nevertheless, spinal angiography is the gold standard and should be taken into consideration where AVF is highly suspected [4, 5, 12]. When these two lesions coexist, special attention should be

given on taking the right decision in order to avoid misinterpretations of MRI findings. This regular error documented by multiple authors in literature will attribute the authorship of the symptoms to the spinal dysraphism [5, 7, 11]. In our opinion any adult patient with spinal dysraphism affected of progressive onset of neurological deficits presenting or not indirect AVF radiological findings in the MRI, should undergo spinal angiography in order to rule out the coexistence of a vascular malformation. Przepiórka et al. described factors such as obesity, aortic aneurysm and aortic atherosclerosis in order to explain the false negatives in spinal angiography [7].

The term of multiple spinal AVFs has been generally used to describe more than one AVF. Inside this group, synchronous AVFs represent those lesions happening simultaneously in different locations. They suppose a rare entity with 19 cases described in literature and a documented 2% rate of occurrence [9] (Table 2). Nevertheless, due to lack of spinal supraselective multilevel angiographies that are normally completed during the examination its frequency can be underestimated and can be wrongly classified as false metachronous when they are discovered in a second angiography. During regular practice, once the AVF is identified further injections of neighbored supra and infrasegmental arteries are not normally performed because the procedure is not exempt from risks [2, 9]. True metachronous AVFs occur even more rarely with only 11 cases described in literature (Table 3). They are defined as those lesions ubicated in a different location which are diagnosed in a temporal gap after the first AVF has been treated and the patient has temporally improved [1, 2].

The most accepted theory about multiple spinal AVFs is based in the occlusive and hypertensive events happening in the venous system in the presence of a single AVF which would promote the overexpression of angiogenic growth factors and create an excellent atmosphere for new AVFs. In other words, the presence of a single AVF can promote de development of a new fistula during time. [8]. According to this theory, in presence of multiple AVFs the existence of a dominant "high-flow" AVF which is clinically manifested can hide and overlook the other(s) AVF(s) as "slow-flow" lesion(s). It is reported that in presence of AVFs a steal phenomenon takes place in the dural microvasculature with compensatory vasodilatation. This compensation enables to preserve dural perfusion in this situation and opens arteriovenous physiologic shunts within de dura and indirectly between the AVFs [3, 18]. When the AVF is treated and the arterial pressure increases, the flow is shifted from arteriovenous channels within the dura to the previous "slow-flow" AVF. Subsequently, once the first "high-flow" AVF is closed the other AVF is straightly enlarged and manifested with new neurological deterioration [2]. In our case a new AVF was manifested 4 months after the treatment of the two first AVFs and even if the first spinal angiography did not disclose the presence of that new left L2 AVF we can't affirm that it was not present as a "slow-flow" AVF by the time. Nevertheless and according to the generally accepted definition of what true metachronous AVFs are, our third AVF is fully considered as metachronous due to the lack of its presence in the first angiogram and the important neurological improvement which took place after the first treatment. The absence of the third AVF diagnosis during the first angiography suggests the following possibilities: One, the third fistula was de novo and created due to venous hypertension and thrombosis. Two, the third fistula was too small and enlarged due to increased explained shunting after first fistula treatment. Three, the angiography was not completed in the level of the feeders of the third fistula [3].

Overall, 20 well-documented cases of spina bifida and AVF are described in literature. Among these group only 4 cases of multiple AVF has been described. As a far as concerned no one has reported a case of this doble coincidence and the concomitant presence of multiple synchronous and metachronous AVFs in the same patient.

The main goal of the treatment is to reach occlusion in order to normalize the venous pressure and reduce its effects to the spinal cord. Endovascular embolization is usually used as a first step and initial treatment as it represents a less invasive treatment [2, 3]. However, the success rate of endovascular treatment described in literature for AVF is among 70-90% in comparison to the 98% documented for open surgery [2, 3, 8, 19, 20]. Recent evidence enforces that open surgical approach and AVF disconnection is preferred and considered a safe, effective and permanent procedure as it is related with lower recanalization rates and occurrence of a second AVF [2, 3, 8, 19, 20]. In presence of single or multiple synchronous AVFs and dual coexistence with spinal dysraphism as in our case, the possibility of performing simultaneous treatment in one-step surgery for both entities seems to be reasonable, safe and definitive [9, 12].

Table 1: Overview the 20 cases of the double association between dysraphism and spinal dural arteriovenous fistulas.

Authors	Age/ Gender	Previous surgery	Lipoma type	Level	AVF type	Feeder	Embo lization	Syrgycal operation	Results
1. M. Djindjan, 1989	53/Male	No	Caudal	S2S3	Dural AVF	M, Lt LSA	Yes	Yes	Improved
2. M. Konig, 1999	50/Male	No	Transitional	L5	Dural AVF	Lt L3 LA	No	Yes	Unknown
3. K. Rajeav, 2005	44/Female	No	Dorsal type	L1L2	Dural AVF	Lt L1 LA	No	Yes	Improved
4. C. Erdogan, 2007	40/Male	No	Dorsal type	L3	Dural AVF	Rt L2	Yes	Yes	Complete recovery
5. M. Sato, 2013	72/Male	No	Dorsal type	L3	Dural AVF	Lt L3 LA, Rt L2 LA	Yes	No	Improved
6. S.B. Mavani, 2014	29/Male	Yes	Transitional	L5S1	Dural AVF	Lt L4 LA	No	Yes	Improved
7. K.M. Krisht, 2015	58/Female	Yes	Dorsal type	L4L5	Dural AVF	Bilateral S2 LA	Yes	Yes	Improved
8. Y. Horiuchi, 2016	51/Male	No	Dorsal type	L5	AVF in lipoma	Rt LSA	Yes	Yes	Improved
9. Y. Horiuchi, 2016	53/Male	No	Dorsal type	L2L3	AVF in lipoma	Rt L2 LA	Yes	Yes	Improved
10. E. Giordan, 2017	46/Male	No	Unknown	S3S4	Dural AVF	Lt S4	Yes	No	Unchanged
11. E. Giordan, 2017	64/Male	No	Unknown	Sacral	Dural AVF	SA	Yes	No	Improved
12. E. Giordan, 2017	44/Female	Yes	Unknown	L4L5	Dural AVF	SA	Yes	No	Worsened
13. E. Giordan, 2017	65/Female	Yes	Unknown	Unknown	Dural AVF	Rt SA	Yes	No	Improved
14. E. Giordan, 2017	57/Male	Yes	Unknown	Unknown	Dural AVF	SA	Yes	No	Not defined
15. E. Giordan, 2017	64/Female	Yes	Unknown	Unknown	Dural AVF	SA	Yes	No	Improved
16. G. Talenti, 2017	19/Male	Yes	Dorsal type	L2L3	Dural AVF	Rt L5 LA, Lt S2 SA	Yes	No	Improved
17. G. Talenti, 2017	53/Male	Yes	Meningocele	L5S1	Dural AVF	Rt S1 SA	Yes	Yes	Unchanged
18. L. Przepiórka, 2018	30/Female	No	Caudal	S2S3	Dural AVF	Lt S3 SA	No	Yes	Complete recovery
19. L. Przepiórka, 2018	33/Male	No	Caudal	S2S3	Dural AVF	Lt and Rt S2S3 SA	Yes	Yes	Improved
20. A. Whitaker-Lea, 2018	57/Female	No	Unknown	Unknown	Dural AVF	Lt SA	Yes	No	Improved
21. Flor-Goikoetxea, 2021	37/Male	No	Dorsal type	L2L3	Dural AVF	Rt L4 LA// Lt L2, L3 LA	Yes	Yes	Improved

Table 2: Overview the 19 cases of multiple synchronous spinal dural arteriovenous fistulas reported in literature.

Authors	Level	Treatment
1. Thiebot, 1986.	Rt T4, Lt T12, Rt L4 and Rt L5	No treatment
2. Pierot, 1993.	Lt T6 and Rt T8	Embolization/surgery
3. Pierot, 1993	Rt T8 and Lt T9	Surgery
4. Chaloupka, 1995.	Lt T9 and L1	Embolization/surgery
5. Dam-Hieu, 2001	Rt T6 y Rt L1	Surgery
6. Krings, 2004	Rt L1 and Lt L2	Surgery
7. El-Serwi, 2006	Rt T5 and T7	Embolization
8. Cenzato, 2007	Rt T5 and Lt T6	Surgery
9. Shankar, 2011	Lt C1,C2 and C6	Incomplete embolization
10. Oshita, 2011	Lt C1 and Rt C1	Surgery
11. Hanakita, 2012	Rt T7 and Lt T12	Surgery
12. Ge, 2013	Lt T12 and Lt L1	Embolization/surgery
14. Hetts, 2013	Lt C5-C6 and Rt C5-C6	Surgery
15. Jablawi, 2018	Lt T8 and Rt T9	Surgery
18. Jablawi, 2018	Lt T7 and Rt T7	Surgery
19. Cannizzaro, 2019	Rt T10 and Lt T11	Surgery
20. Present case	Rt L4 and Lt L3	Embolization

Table 3: Overview the 11 cases of multiple metachronous spinal dural arteriovenous fistulas reported in literature.

Author	Level	Treatment
1. Barnwell/1991	Rt C2 and R tC7	Surgery
2. Van Dijik/2001	T5 and T9 (N.D)	(N.D)
3. Ling/2005	T11 and T12 (ND)	Embolization/surgery
4. Sugawara/2005	Rt T6 and Lt L1	Surgery
5. Rizvi/2006	Rt T9 and Lt L1	Embolization/surgery
6. Dagar/2010	Lt T12 and Rt L1	Surgery
7. Avecillas-Chasin/2015	Rt C1 and Left VA	Surgery
8. Avecillas.Chasin/2015	Rt T7 and T12	Embolization/surgery
9. Kaku/2017	Rt C1 and Rt S2	Surgery
10. Jablawi/2018	Rt L1 and Lt L2	Surgery
11. Ren/2019	Lt T10 and Rt L1	Surgery
12. Present case	Rt L4/Lt L3 and Left L2	Surgery

5. Conclusions

The concurrence of multiple synchronous and metachronous AVFs and spinal dysraphism at the same level is exceptionally rare. The exponentially growing number of published cases in literature strongly support that this relationship is not a coincidence. In terms of pathophysiology most authors underline the potential shared congenital and acquired origin as the mixed mechanism of the development of AVF.

AVF should be strongly suspected in adults with previous history of spinal dysraphism and progressive onset of neurological deficits by performing gold standard spinal angiography to rule out its existence.

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