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Spinal arteriovenous shunts presenting as intracranial subarachnoid haemorrhage

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Abstract *Background* In approximately 5% of patients with intracranial subarachnoid haemorrhage (SAH), the cause is another than a ruptured aneurysm or perimesencephalic haemorrhage. One of these causes is a spinal arteriovenous shunt (SAVS). The aim of this study was to investigate the characteristics of patients with SAVS who present with intracranial SAH without symptoms and signs suggesting a spinal cause. *Methods* We systematically reviewed the literature and searched the SAH database of the University Medical Center Utrecht, The Netherlands, for patients with SAVS presenting with intracranial SAH and studied the characteristics of patients with SAVS whose clinical presentation mimicked intracranial SAH caused by rupture of a saccular aneurysm. *Results* Thirty-five patients were identified after a review of the literature. In our SAH database, comprising 2142 patients included in the period 1985–2004, we found one patient (0.05%, 95 % CI 0.006–0.3%). SAH due to SAVS occurred at any age (4–72 years). The SAVS was located at the craniocervical junction in 14 patients, at the cervical level in 11, and at the thoracolumbar level in the remaining 11 patients. The majority of patients (n = 26, 72%) had no disabling deficits at discharge or follow-up. *Conclusion* Rupture of a SAVS presenting as intracranial SAH is rare and can occur at any age. The SAVS can be located not only at the craniocervical junction or cervical level but also in the thoracolumbar region. Most patients with SAVS presenting as intracranial SAH have a good recovery.

Key words subarachnoid haemorrhage · spinal arteriovenous shunt

Abbreviations AVF: Arteriovenous fistula, AVM: Arteriovenous malformation, CCJ: Craniocervical junction, GOS: Glasgow Outcome Scale, IVH: Intraventricular haemorrhage, LP: Lumbar puncture, SAH: Subarachnoid haemorrhage, SAVS: Spinal arteriovenous shunt

Introduction

In approximately 15% of patients with intracranial subarachnoid haemorrhage (SAH), the cause of the haemorrhage is another than a ruptured intracranial saccular aneurysm. Two-thirds of these patients, thus comprising 10% of all patients with intracranial SAH, have a non-aneurysmal perimesencephalic haemorrhage. The remaining 5% of intracranial SAH

is caused by a variety of rare conditions such as a (transmural) arterial dissection, a cerebral arteriovenous malformation, a cranial dural arteriovenous fistula, a mycotic/septic aneurysm, pituitary apoplexy, cocaine abuse, sickle cell disease, a coagulation disorder or trauma [1, 2]. In addition, spinal arteriovenous shunts (SAVS), in particular if localized in the cervical region, have been reported to present with symptoms and signs suggesting an intracranial cause of the SAH [1, 2].

Most commonly, SAVS present with gradually worsening sensory disturbances, diffuse back and muscle pain, weakness and sphincter disturbances. Acute onset of symptoms is mostly attributed to spinal haemorrhage either into the subarachnoid space or intramedullary and rarely to venous thrombosis [3, 4]. SAVS can be divided into arteriovenous fistulas (AVF) and arteriovenous malformations (AVM). Spinal AVF commonly presents with signs and symptoms of progressive myelopathy. Spinal AVM may also present with pain, acute myelopathy, or radiculopathy [5]. Spinal SAH is reported in approximately half of symptomatic spinal cord AVM [6, 7] and is frequently accompanied by intracranial signs and symptoms [8].

The aim of this study was to investigate the clinical characteristics and type and location of the SAVS of patients with SAVS who present with symptoms and signs suggesting an intracranial and not a spinal cause of the SAH. We also aimed to assess the proportion of patients with intracranial SAH in whom the cause is a SAVS.

Methods

Review of the literature

To identify patients with intracranial SAH caused by SAVS we performed a literature search using PubMed (up to December 2005) with the following keywords: spinal arteriovenous malformation(s), spinal arteriovenous shunt(s), spinal vascular malformation(s) and subarachnoid haemorrhage. Additional articles were found by searching the reference lists of relevant articles.

We included all articles reporting patients diagnosed with intracranial SAH caused by a SAVS with extracranial arterial supply, without signs and symptoms directly suggestive of spinal pathology. Patients were included when a diagnosis of intracranial SAH was reported. Preferably the diagnosis was confirmed on CT but patients with confirmation of SAH by cerebrospinal fluid investigation only or patients, in whom the details on how the diagnosis of intracranial SAH was reached were lacking, were not excluded. Articles in languages other than English were excluded.

Two authors (J.v.B, D.C.G.S) independently reviewed and extracted the following data on eligible patients: demographic characteristics, method of diagnosis of SAH, medical history, presenting symptoms, findings at neurological examination on admission, neuro-imaging, angio-architectural features of SAVS, classification of SAVS (according to Spetzler et al. [5]), treatment, clinical and radiological follow-up (including obliteration after treatment), and clinical outcome, which we aimed to classify according to the Glasgow Outcome Scale (GOS) [9]. In case of disagreement between these two authors, extracted items were reviewed together to reach consensus. If necessary, a third author (C.J.M.K.) resolved the disagreement between the first two authors. The included cases were subdivided into three subgroups: craniocervical junction SAVS (CCJ, between the foramen magnum and C2), cervical SAVS (C3-C8), and thoracolumbar SAVS.

Search SAH database

The records of all patients registered in the SAH database of the University Medical Center Utrecht from January 1985 to January 2005 were reviewed to find additional cases. The same inclusion and exclusion criteria as for inclusion of cases reported in the literature were applied. The method of data-extraction was identical to that for the patients reported in the literature.

Data analysis

The data were analyzed using SPSS for Windows, version 12.0.1 (SPSS Inc.). We used the Students T-test to compare mean ages between the different subgroups of patients. We used proportions with corresponding 95% confidence intervals (CI) to assess the estimated prevalence of SAVS among patients registered in our SAH database and to compare clinical and radiological characteristics of the patient subgroups.

Results

The literature search yielded 35 patients reported in 28 articles. In the SAH database, consisting of 2142 patients with SAH in the studied period, we found one patient (0.05%, 95% CI 0.006–0.3%). Of the 2142 patients, 1740 (81.2%) patients were diagnosed with an aneurysmal SAH, 149 (7.0%) with a perimesencephalic haemorrhage, and 130 (6.1%) with another cause, including the one SAVS. In 123 (5.7%) patients no definite cause was identified. All patients underwent four-vessel angiography if CT angiography demonstrated no abnormalities but we have not systematically searched for SAVS by MRI screening in every patient with a negative angiogram.

A summary of the 36 patients with intracranial SAH caused by SAVS is presented in Table 1. In 14 of the 36 patients (39%), the SAVS was located at the CCJ, in 11 patients (30.5%) at the cervical level, and in another 11 (30.5%) at the thoracolumbar level. In only one of the 11 patients with a thoracolumbar SAVS, the malformation was located at the midthoracic level.

Twenty-two patients (61%) were men. Mean age of patients at presentation was 36 years (range 4–72 years). Patients with a SAVS at the CCJ presented at older age than patients with a SAVS in the cervical or thoracolumbar region (mean difference, 33 years; 95% CI 21–44 years; p-value < 0.0001). The delay between the (first) SAH and the diagnosis of a SAVS

Table 1 Summary of characteristics of 36 patients with intracranial SAH caused by SAVS

First author (year of publication)	Gender	Age*	n of SAH	Delay[a]	Location SAVS	Classification SAVS
Craniocervical SAVS (n = 14)						
Aviv (2004) [10]	M	57	1	No delay	C1	Unknown
	F	53	1	No delay	C2	Unknown
Do (1999) [11]	M	50	1	No delay	C1	Unknown
Endo (2001) [12]	M	70	1	No delay	C1	Unknown
Hashimoto (2000) [13]	F	66	1	No delay	C1	Dorsal intradural AVF
Hosoda (1994) [14]	M	69	1	No delay	CCJ	Intramedullary AVM
	M	59	1	No delay	CCJ	Intramedullary AVM
	M	70	1	No delay	CCJ	Intramedullary AVM
Kai (2005) [15]	M	54	1	No delay	CCJ	Ventral intradural AVF
Kinouchi (1998) [16]	M	68	1	No delay	C1	Unknown
Markert (1996) [17]	F	72	1	No delay	C1-C2	Ventral intradural AVF
Morris (1960) [18]	M	34	2	No delay	C1-C2	Unknown
Yaşargil (1975) [19]	M	37	1	No delay	C1-C2	Unknown
	F	31	12	9	C1-C2	Unknown
Mean (SD)		56 (14)				
Cervical SAVS (n = 11)						
Bassuk (2003) [20]	F	13	1	No delay	C4	Extra-intradural AVM
Halbach (1993) [21]	F	11	1	No delay	Midcervical	Ventral intradural AVF
Henson (1956) [22]	M	13	2	2	C1-4	Unknown
Hida (2002) [23]	F	62	1	No delay	C5	Ventral intradural AVF
Höök (1958) [24]	F	25	2	9	C4-6	Unknown
Morimoto (1992) [25]	M	61	1	No delay	C5	Unknown
Odom(1962) [26]	F	11	1	No delay	C3-7	Unknown
Willinksy (1990) [27]	M	36	1	No delay	C8	Dorsal intradural AVM
Yaşargil (1975) [19]	M	20	9	15	C3	Unknown
	M	21	1	4	C2-5	Unknown
	F	18	4	7	C5-7	Unknown
Mean (SD)		26 (19)				
Thoracolumbar SAVS (n = 11)						
Clark (1995) [28]	M	4	1	No delay	T9-L2	Dorsal intradural AVF
Cogen (1983) [29]	M	7	3	2	T9-12	Intramedullary AVM
Koch (2004) [30][b]	F	46	1	No delay	L4	Dorsal intradural AVF
Maggioni (1995) [31]	F	23	1	No delay	T10-12	Intramedullary AVM
Mandzia (1999) [32]	M	9	1	No delay	T12	Conus medullaris AVM
Parkinson (1977) [33][c]	M	47	2	5	T12	Unknown
Rosenow (2000) [34]	M	27	1	No delay	T10-11	Ventral intradural AVF
van Santbrink (2003) [35]	M	38	1	No delay	L1	Conus medullaris AVM
Wakai (1992) [36]	M	6	2	2	C7-T2	Ventral intradural AVM
Williams (1991) [37]	F	16	4	3	T8	Intramedullary AVM
This study (2006)	F	6	1	No delay	T12-L3	Extra-intradural AVM
Mean (SD)		21 (17)				

AVF Arteriovenous fistula, *AVM* Arteriovenous malformation, *CCJ* Craniocervical junction, *F* Female, *M* Male, *n* Number, *SAH* Subarachnoid haemorrhage, *SAVS* Spinal arteriovenous malformation. *With first symptoms of SAH; [a] Delay between first SAH and diagnosis of SAVS (in years); [b] In this patient, a posterior communicating artery aneurysm was initially thought to be the cause of her SAH. During the operation, the surgeon was not convinced that the aneurysm had ruptured and caused the symptoms. A subsequent MRI showed a lumbar SAVS surrounded by a subarachnoid blood clot and was confirmed after spinal angiography [30]; [c] In this patient, intracranial SAH was first attributed to a temporal AVM. After angiographically confirmed total excision of the AVM, the patient presented with another intracranial SAH 4 years later. Four-vessel angiography revealed no intracranial vascular malformation but a subsequent myelogram demonstrated a thoracolumbar SAVS [33]. Eight patients presenting with SAH due to a SAVS were not included in this table because of symptoms that at first presentation immediately indicated a cause with spinal location: presentation with a paraparesis [38, 39], quadriparesis, and sensory disturbances below the midcervical level within a few hours after sudden severe headache [19, 40], and back pain as predominant clinical symptom [41, 42]. An additional patient was diagnosed with a spinal AVM in 1967 when he presented with a bilateral foot drop, increased lower extremity deep tendon reflexes with extensor plantar responses, and a sensory impairment below T8 level. Five years later, a cerebral AVM and a large cervical AVM were diagnosed on angiography after he presented with a two-month history of headaches and right leg pain. After another two years, he presented with an acute SAH that could be attributed to either AVM [43]

varied from 0 to 15 years. A delayed diagnosis had occurred mostly in cervical (n = 5; delay of 2–15 years) and thoracolumbar SAVS (n = 4; delay of 2–5 years). Twenty-six patients (72%) had only one episode of SAH, the remaining ten patients had two or more episodes (range 2–12 SAH). In all ten patients with two or more episodes, the diagnosis was made only after a repeated episode. The presenting symptoms, findings on examination, CT findings, and treatment results of all 36 patients are listed in Table 2.

Clinical presentation

Most patients (n = 27, 75%) presented with headache. In two of the remaining nine patients, no clinical details about the first SAH were reported but these two patients did present with headache after subsequent SAH [19, 22]. In the other seven patients, primarily patients with cervical or thoracolumbar SAVS, there was no information on whether or not patients also had headache at the time of presentation. Two of these seven patients presented with impaired consciousness [28, 32]. In one of them, generalized tonic seizures, abnormal breathing pattern, and bilateral papiledema with retinal haemorrhages were reported [28]. In one other patient, who presented with a mild hemiparesis, CT of the brain showed SAH [27, 29]. The reported signs and symptoms that may have added to the diagnosis of intracranial SAH in another two patients for whom there was no information on the presence or absence of headache, were vomiting and dizziness in one patient [14] and nuchal rigidity in another [29]. In two patients no further information was available [19].

A period of unresponsiveness was found in eight patients (22%), three of whom had a thoracolumbar SAVS. Back pain associated with the symptoms and signs suggesting an intracranial cause was found only in patients with a thoracolumbar SAVS; it occurred in four (36%) of the patients with thoracolumbar SAVS. Neck pain was reported in nine patients (25%), four of whom had thoracolumbar SAVS. In eight of these nine patients, the neck pain was associated with headache. In the ninth patient, a 13-year-old boy, acute pain in the neck was the only symptom during the first episode of SAH. At the second episode he also had headache and vomited. At this second episode, a cervical vascular malformation was diagnosed after vertebral angiography [22]. Only one of the 11 patients with a cervical SAVS complained of pain between the shoulder blades after this patient had a

Table 2 Clinical characteristics, diagnostic tests, and treatment in 36 patients presenting with SAH due to SAVS

	All patients		CCJ SAVS			Cervical SAVS			Thoracolumbar SAVS		
	n = 36	%	n = 14 Present	Absent	Unknown	n = 11 Present	Absent	Unknown	n = 11 Present	Absent	Unknown
*Symptoms**											
Headache	27[a]	75[a]	13	–	1	6	–	5[a]	8	–	3
Nausea	9	25	4	–	10	1	–	10	4	–	7
Vomiting	11[a]	31[a]	4	–	10	3	–	8[a]	4	1	6
Unresponsiveness	8	22	4	1	9	1	1	9	3	1	7
Neck pain	9	25	2	1	11	3	–	8	4	1	6
Back pain	4	11	–	1	13	–	1	10	4	–	7
Neurological examination											
Impaired consciousness	4	11	2	4	8	–	1	10	2	1	8
Nuchal rigidity	13[a]	36[a]	5	–	9	2	1	8[a]	6	1	4
Focal neurological deficit	6[a]	17[a]	1	8	5[a]	3	3	5[a]	2	5	4
Brain CT											
Performed	25	69%	11	3 NA	–	4	6 NA	1	10	1 NA	–
Confirming SAH	17	47%	9			2			6		
Blood distribution (CT)											
Basal cisterns	7	19	3			–			4		
Perimesencephalic	1	3	1			–			–		
Intraventricular	10	28	6			–			4		
Third ventricle	3	8	1			–			2		
Fourth ventricle	9	25	5			–			4		
Cisterna magna	2	6	1			1			–		
SAH, not specified	6	17	3			1			2		
Negative CT	8	22	2			2			4		
Treatment SAVS											
Surgery	23	64	12			6			5		
Embolization	4	11	–			1			3		
Embolization and surgery	3	8	–			1			2		
Radiation	1	3	–			1			–		
None	5	14	2			2			1		
Results treatment SAVS	n = 31										
Partial obliteration	4	13	–			2			2		
Complete obliteration	17	55	9			3			5		

**CCJ* Craniocervical junction, *NA*, Not available, *SAH* Subarachnoid haemorrhage, *SAVS* Spinal arteriovenous malformation, *At first presentation, [a] For three patients details of their first SAH were not described but information about subsequent SAH were documented [19, 22]

second SAH [24]. Two patients (6%) had a history of transient spinal cord signs before the SAH; one of these patients had a transient paraplegia 6 years before SAH, and the other had a history of quadriparesis 11 years before the SAH (diagnosed as Pertussis and Poliomyelitis) [26, 37]. One patient mentioned, after further questioning, difficulty with urination in the year before presentation with a SAH [34].

Six patients had neurological deficits at the time of the first episode of SAH. A hemiparesis was present in three patients [14, 24, 27]. One patient presented with a weakness of all extremities, but more pronounced on one side [26]. Two patients presented with a mild weakness in one leg only [34, 37]. The sixth patient had a peripheral facial nerve palsy [12]. Papiledema [20, 28] and retinal haemorrhages [28] were found in three patients (9%). Presence of nuchal rigidity was reported in two patients (18%) with cervical SAVS, in five patients (36%) with CCJ SAVS, and in six patients (55%) with a thoracolumbar SAVS from a total of 15 patients in whom information on the presence or absence of nuchal rigidity was reported.

Seven patients developed neurological deficits after their first SAH but before diagnosis [19, 27, 36]. In four patients, this was after additional episodes of SAH [19, 22]. The signs and symptoms included quadriparesis [19, 27], paraparesis with weakness of one arm [19], hemiparesis [19], paresis of one leg [36], sphincter [22, 27] and sensory [19, 36] disturbances, and decreased unilateral abdominal reflexes [22].

Diagnostic procedures and neuro-imaging

A brain CT was done in 25 (69%) of the 36 patients, and showed SAH in 17 (47%). Details about the delay between initial symptoms and timing of CT were not mentioned. In 12 patients (34%), the diagnosis of SAH was based upon a positive lumbar puncture. In seven (20%) patients, the diagnostic method was not mentioned. In four of them there was headache or pain in the neck, and in three patients details on the clinical presentation were not given. Six of these seven patients presented with a first episode of SAH in the fifties [22], sixties [19] or early seventies [33] of the last century, when CT scanning was not available. The seventh patient presented with severe headache from SAH, but a four-vessel cerebral angiography showed no abnormalities, and subsequent cervical spinal MRI scan revealed a vascular malformation [21].

The distribution of blood on the brain CT is summarized in Table 2. Intracranial SAH on CT was frequently seen in CCJ SAVS (in nine of 11 patients in whom a CT was performed), but also in the majority (six of ten patients in whom a CT was performed) of patients with thoracolumbar SAVS and in half of patients with a cervical SAVS (in two of four patients in whom a CT was performed).

In 13 of the 14 patients with a CCJ SAVS cerebral angiography revealed the diagnosis, in the remaining patient after spinal angiography. The diagnosis of a cervical SAVS was made after cerebral angiogram in five of the 11 patients, on a diagnostic laminectomy [19, 27] in three, on spinal MRI in two [20, 21], and after myelography in one patient [26]. Three CCJ SAVS [10, 11, 18] and three cervical SAVS [19, 24, 25] were not diagnosed directly after the first cerebral angiogram because initially not all four cerebropetal vessels were catheterized selectively. Of the 11 patients with thoracolumbar SAVS, seven patients were diagnosed with MRI, two after spinal angiography, and two with myelography.

Treatment and clinical outcome

Patients with a SAVS at the CCJ were treated surgically (86%) or received no treatment to obliterate the SAVS (14%). Embolization was performed only in cervical and thoracolumbar SAVS. Three patients were reported to await treatment and two patients [18, 26] did not receive treatment of the SAVS. Five of the 36 patients (14%) were treated because they developed hydrocephalus. Three patients needed temporary drainage by means of an external ventricular drain [10, 28]. A ventriculoperitoneal drain was placed in one patient [14]. Another patient was treated with acetazolamide to reduce intracranial pressure [20].

Information on outcome was available for 33 patients. Twenty-six patients (79%; 95% CI 61–91%) made a good recovery (GOS 5), four patients remained moderately disabled (GOS 4), and two patients were severely disabled (GOS 3). One patient died of shunt-related complications [10].

Discussion

This study of 36 patients with SAVS presenting with signs and symptoms suggesting an intracranial cause of SAH showed that intracranial SAH from a SAVS can occur at any age and that the SAVS can be located at any level: at the craniocervical junction, the cervical level and at the thoracolumbar level. Most patients with intracranial SAH from a SAVS have a good recovery.

SAVS have been reported as a rare cause of intracranial SAH. However, mainly patients with cervical SAVS have been described as being indistinguishable from patients with an intracranial source

of bleeding [1, 2, 44]. Our search retrieved 11 patients with a thoracolumbar SAVS, emphasizing that SAVS localized at any level of the spinal cord can present with signs and symptoms of intracranial SAH.

We found no SAVS located between the thoracic levels two and eight. Because of the small number of patients included in this study this is most likely due to chance. In general, SAVS frequently occur in the midthoracic region [7, 45]. Dorsal intradural spinal AVF are found mainly in the low thoracic and lumbar regions in contrast to other types of SAVS that are spread more equally along the cord [7, 46, 47]. In this study, two dorsal intradural spinal AVF were located in the thoracolumbar region [28, 30], but in two other patients dorsal intradural spinal AVF were situated in the cervical region [13, 27].

The pathological mechanism of intracranial SAH from a SAVS remains unclarified. The most straightforward mechanism is migration or extension of subarachnoid blood from the spinal to the intracranial level [26, 28, 31]. Haemorrhage may be caused by venous hypertension when arterialized blood flows via the medullary vein to the valveless coronal venous plexus and radial vein [11, 25]. Another hypothesis suggests that the vein around the midbrain is compressed or stretched by the tentorial incisura when, eg., physical exercise elevates the ICP, which then leads to aggravation of venous hypertension with subsequent rupture of the vein [13]. In patients with intracranial drainage of their SAVS, the relatively fast venous flow may cause formation of a varix on the draining vessel, which may result in intracranial SAH after rupture [16]. Ascending venous drainage was associated with an increased risk of SAH in six patients with CCJ perimedullary and dural AVF [15]. In this light, it is not surprising that in patients with SAVS also other cranial symptoms and signs than those suggestive of intracranial SAH caused by rupture of a saccular aneurysm have been reported, such as intermittent double vision, slurred speech, and nystagmus [8].

A limitation of this study is that in only about half of the reported patients with SAVS presenting with intracranial SAH the diagnosis was confirmed on brain CT. However, in all 36 patients the authors were convinced of the diagnosis of intracranial SAH and at first instance searched for a saccular aneurysm.

It remains questionable whether in patients with SAH without an intracranial source of the haemorrhage SAVS should be searched for, since intracranial SAH caused by SAVS is very rare. Four-vessel angiography can detect a CCJ or cervical SAVS and should always be performed before investigating the spinal cord. MRI screening of the spine in all patients with (repeated) negative angiograms will probably have a low yield and be very expensive. In a cohort of 15 patients with an aneurysmal pattern of SAH and three negative four-vessel angiograms, but no imaging of the spine, no episodes of new episodes of SAH occurred during an average follow-up of 65 months [48]. We suggest that in patients with an intracranial SAH without identifiable cause, close attention should be given to sometimes subtle clues in the history and examination (such as back pain, pain between shoulder blades, and difficulty with urination) that may point to a spinal cause of the SAH. A delayed diagnosis can have negative implications for patients, because they may develop new or progressive neurological deficits that may or may not be related to new episodes of SAH. In patients with multiple episodes of SAH and repeated negative cerebral four-vessel angiograms, further investigations (e.g., MRI) should probably be considered to exclude a SAVS as a rare but possible cause, even in the absence of symptoms or signs of medullary or spinal root involvement. In those rare cases the full length of the spinal cord should be investigated.

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