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Case Report

Rare association of secondary superficial siderosis caused by a fourth ventricle hemorrhagic ependymoma mimicking a cavernoma: Case report and literature review

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Abstract

Background: The association of a hemorrhagic tumor with secondary superficial siderosis (SS) is a relatively rare although well described phenomenon.

Case Description: We present the case report of a 35-year-old male with a history of drowsiness, hypoacusia, drop attacks, and multidirectional nystagmus during the last 2 months, who presented with acute obstructive hydrocephalus caused by a fourth ventricle mass displaying radiological signs of repeated intra and extratumoral hemorrhage with SS. He underwent gross surgical removal of the solid component of the tumor. Microscopic examination revealed an ependymoma with atypical features, including prominent angiomatous formations and internal chronic hemorrhages with hemosiderin deposits, resembling a cavernoma. The scarce tumoral component, which extended around these cavernous vessels, lacked the gross typical features of fibrillary stroma or perivascular pseudorosettes.

Conclusion: To our knowledge, including the present case, there are 45 published reports of tumors associating secondary SS. Besides ependymoma, no other hemorrhagic lesion, tumoral or vascular, has been previously published associating a fourth ventricle location with secondary SS. The present case represents the fifth with this finding, and we strongly suggest ependymoma as a presumptive diagnosis when this rare association is encountered. In addition, this appears to be the first case reported in the scientific literature of a hemorrhagic fourth ventricle ependymoma mimicking both, radiologically and histologically, a cavernous malformation.

Key Words: Cerebral ventricle neoplasms, fourth ventricle, subarachnoid hemorrhage

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INTRODUCTION

Superficial siderosis (SS) of the central nervous system (CNS) is a rare condition caused by chronic bleeding in the subarachnoid compartment. The source of bleeding remains unknown in up to 35% of cases.[31]

The pathologic changes associated with SS have been previously described. Macroscopically, the

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leptomeninges and superficial CNS parenchyma, as well as the subependymal lining throughout the neuroaxis, present a dark, brownish discoloration. Microscopically, extensive hemosiderin deposition can be found in the leptomeninges, as well as in the subpial and subependymal regions. In addition, the leptomeninges are thickened, and varying degrees of neuronal loss, reactive gliosis, and demyelination can be found. In the cerebellum, the superficial folia are almost always involved with loss of Purkinje cells and Bergmann gliosis. Particularly dense hemosiderin deposition can be found in cranial nerve VIII, and to a lesser extent in cranial nerves I and II; these findings are often associated with demyelination and atrophy.[38]

We present a case of SS caused by an ependymoma with unique radiological and pathological features, and include a revision of the current scientific literature concerning this association.

CASE REPORT

History

A 35-year-old male was brought to the emergency department after suffering repeated episodes of drop attacks. He was being studied by an ENT specialist because he had complained of drowsiness, gait instability, left neurosensorial hypoacusia, cervicalgia, and multidirectional nystagmus during the last 2 months.

Examination

In the initial neurological examination, he scored 13 in the Glasgow Coma Scale (GCS) (E: 3; V: 4; M: 6), and presented mild papilledema, horizontal-rotatory nystagmus, and instability.

Imaging and initial management

Emergent computed tomography (CT) [Figure 1a] revealed a heterogeneous fourth ventricle mass causing acute obstructive hydrocephalus (HCP). An emergent insertion of an external ventricular drainage (EVD) was carried out and the patient experienced a complete recovery of his level of consciousness. A brain magnetic resonance imaging (MRI) was then obtained [Figure 1b-d], which confirmed the presence of a fourth ventricle lesion displaying signs of both intra and extratumoral hemorrhage with SS. These findings suggested a radiological diagnosis of myxopapillary ependymoma versus cavernoma. The panspinal MRI excluded the presence of additional lesions.

Operation

The tumor was resected through a bilateral telovelar approach with neurophysiological monitoring [Figure 2]. The EVD was withdrawn on the second postoperative day.

Pathological findings

Histologically, the lesion presented a capsule and was mainly composed of great cavernous vessels, with areas of hemorrhages and hemosiderin deposits, thus resembling a cavernous malformation. A scarce tumoral component was found in the periphery of the lesion, which was frequently arranged around hyalinized vessels. Tumoral cells showed an eosinophilic, unclearly delimited cytoplasm and pleomorphic nuclei, with finely granular chromatin and small nucleoli. These tumor cells were glial fibrillary acidic protein (GFAP) positive, and they showed dot-like intracytoplasmic epithelial membrane antigen (EMA) immunoreactivity [Figure 3].

Postoperative course

The patient experienced an almost complete recovery from his initial symptoms in the following month, although he still had persistent neurosensorial hypoacusia and nystagmus. Serial follow-up MRI performed 3, 6, and 12 months after surgery showed compensated triventricular dilation and a stable tumoral remnant on the rostral part of the fourth ventricle; however, the patient refused further treatment.

DISCUSSION

Approximately 0.9–11% of spontaneous intracranial hematomas are produced by brain neoplasms. Tumors presenting as hemorrhagic lesions have a relatively low frequency (2–-11%), and up to 42% remain clinically silent at the time of bleeding.[33,62] Among tumoral hemorrhages, three patterns can be distinguished, namely, pure intratumoral (53%), pure extratumoral (intraventricular

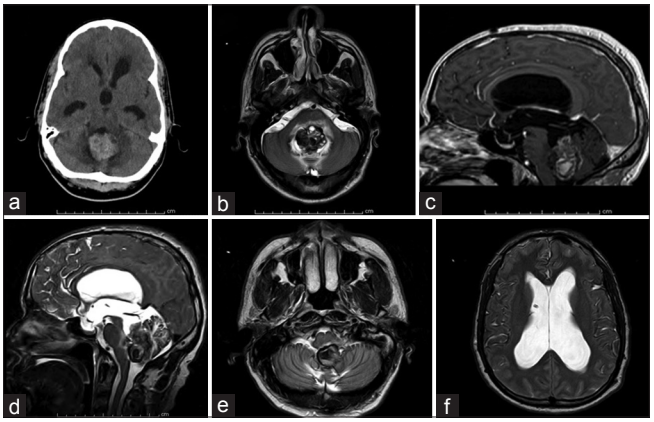

Figure 1: (a) Brain CT showing a 3 × 3× 4 cm 4th ventricle mass, predominantly hyperdense, causing active triventricular dilation. (b) MRI showing a heterogeneous lesion with cystic areas in both T1WI (c) and T2WI (b and d), scarce areas of enhancement (c) and associated edema (b). Linear hypointense signal, in T2WI, along the pial surface/subarachnoid space of the convexity sulci, cerebellar folia, and brainstem and spinal surface, is typical of SS (b, d, e, f). Signs of compensated hydrocephalus are also present (bulging suprasellar cistern, remodellation of the sella turcica) (c and d)

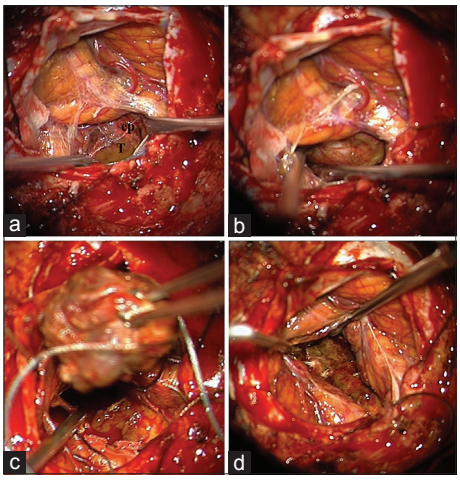

Figure 2: Sequence of photographs obtained during the microsurgical procedure. (a and b) A bilateral telovelar approach was carried out. Notice the brownish colored pial surface of the cerebellum, typical of SS. After evacuation of xanthochromic CSF, a rubbery mass was identified occupying the fourth ventricular chamber. A friable xantochromic material covered both the tumor and the boundaries of the fourth ventricle. Although this material facilitated the definition of a plane of dissection that allowed an en block resection of the tumor (c), it precluded the identification of the anatomical structures of the floor of the fourth ventricle (d), except clear CSF gushing forth from the aqueduct

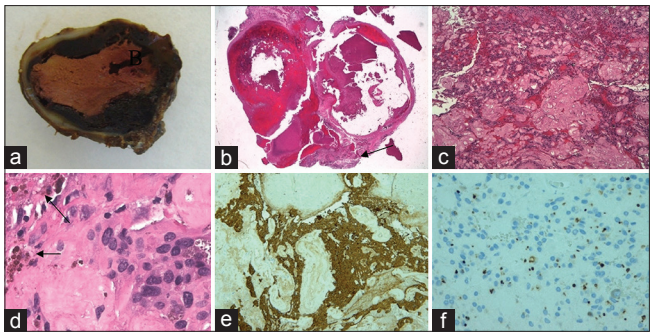

Figure 3: (a) Macroscopic aspect of the lesion, with a heterogeneous surface delimited by a capsule of variable width, which is irregular on one of its borders, with a brownish content. (b) Great vessels with thickened walls of cavernous appearance and minimal areas of tumor in the periphery of the lesion (arrow) (H and E ×4). (c) Tumor cells arranged around hyalinized vessels and signs of recent hemorrhage (H and E ×10). (d) Hemosiderinic pigment (arrows) and tumoral cells with an eosinophilic unclearly delimited cytoplasm and nuclei with finely granular chromatin and small nucleoli (H and E ×40). (e) Perivascular tumor cells express GFAP. (f) Dot-like intracytoplasmic EMA immunoreactivity in tumoral cells

and/or subarachnoid, 33%) and mixed intra and extratumoral hemorrhage (13%).[4] Intratumoral hemorrhage occurs mainly in highly vascular or malignant neoplasms. Excluding pituitary adenoma, which has been found to have a statistically significant higher frequency, those commonly associated with spontaneous intracranial hemorrhage comprise glial tumors, metastatic tumors, meningioma, and choroid plexus papilloma, the majority of which are described in a supratentorial location. An intracranial hemorrhage originated in a tumor located in the posterior fossa represents a relatively uncommon phenomenon (≤0.4%) with a preference for pediatric patients; the predominant histologies described in this location are pilocytic astrocytoma, medulloblastoma, ependymoma, and melanoma.[27,46,62]

Those tumors which produce repeated extratumoral hemorrhages may lead to SS. This entity, first described by Hamil in 1908,[16] was mostly a postmortem diagnosis until the advent of modern neuroradiological techniques. The first description of its radiological features was performed by Gomori in 1985.[13] Currently, it can be defined as a relatively rare condition in which deposits of hemosiderin accumulate in the subpial layer of the CNS as a consequence of prolonged or recurrent low-grade bleeding into the cerebrospinal fluid, which may lead progressively to irreversible neurological dysfunction. The classic clinical triad consists of sensorineural hearing loss (uni or bilateral), cerebellar ataxia, and myelopathy. The latter, together with cognitive decline, has a propensity to appear in secondary forms of the disease that have progressed over many years. Other symptoms suggestive of arachnoiditis, including neckache, backache, and sciatica, are less frequent. These symptoms are not expected to improve with the treatment of the source of bleeding, as occurred in our patient, and only a slight amelioration has been obtained with the employment of iron chelating agents and radical scavengers.[11,28]

SS is a secondary condition in up to 65% of cases.[31] Including the present case, we have recorded a total of 45 published reports of SS in patients with confirmed, nonoperated, current neoplasms of the CNS [Table 1]. Among these cases, 50% were caused by an ependymal tumor and only 5 were located in the fourth ventricle.[6,10,15,50] In our case, histopathologically, the mass presented a capsule and was mainly composed of great cavernous vessels, with areas of hemorrhages and hemosiderin deposits – a sign of recent and old intratumoral bleeding, respectively – thus microscopically mimicking a cavernoma. The abundant presence of these anomalous vessels was the probable cause of the chronic bleeding, and given its location with free access to the CSF, finally led to the development of SS. A scarce tumoral component was found in the capsule and surrounding these vessels, but lacked areas of fibrillary stroma or perivascular pseudorosettes, which complicated the diagnosis of ependymoma.[40] Supporting this diagnosis were the GFAP expression and the dot-like intracytoplasmic EMA immunoreactivity found in the tumoral cells. The radiological findings suggested a preoperative differential diagnosis which included cavernous malformation and myxopapillary ependymoma. After revising the literature on the association of these pathologies with SS, we have found 13 cases of cavernoma with secondary SS, none of which had a fourth ventricle location,[17,18,24,25,29,30,32,35,42,47,49] and

Table 1: Published reports of current, nonoperated CNS tumors presenting with superficial siderosis

	Reference	Age/ Sex	Presentation		HCP/ Treatment	SOB/Location	Management	Follow-up/Outcome
			SS	Other				
1	Noetzel 1940	47/M	Deafness, dementia	No	NA	Metastasis*/CxMen	NA	NA
2	Rosenthal 1958	27/M	NA	NA	NA	Oligodendroglioma/NA	NA	NA
3	McGee 1962	54/M	Deafness, myelopathy	Incontinence	NA	Ependymoma/Lum	NA	NA
4	Dastur 1962	26/M	Meningismus	H/A, N/V, papilledema, ataxia	Obstructive/ VP	Pinealoma	STR	1d/Died
5	Tomlinson 1964	16/F	Deafness, ataxia, dementia, incontinence	H/A, dysarthria	Yes/No	Ependymoma/LV	Not treated	2y/Died
6	Brahman 1965	NA/F	NA	NA	NA	Astrocytoma/NA	NA	NA
7	Kott 1966	29/F	Deafness, myelopathy, meningismus	H/A, N/V, papilledema, seizures, bilateral VIcp palsy	No	Ependymoma/LV	SR	1m/Died
8	Sherwin 1972	31/M	NA	NA	NA	Meningioma/NA	NA	NA
9	Gomori 1985	32/M	Deafness, tinnitus, nystagmus	Sciatica	Yes/No	Myx ependymoma/ Lum	SR	NA/Sciatica, sphincter dysfunction
10	Koeppen 1988	59/M	Deafness, ataxia, myelopathy	No	No	Ependymoma/Lum	GTR + IChA	NA
11	Parnes 1992	59/M	Deafness, ataxia, myelopathy, tinnitus	No	NA	Ependymoma/Lum	NA	NA
12	Willeit 1992	59/M	Deafness, ataxia, myelopathy	Incontinence	No	Ependymoma/Lum	GTR	NA
13	Shen 1993	16/F	Meningismus	No	No	Myx Ependymoma/ Lum	GTR	NA
14	Mamourian 1993	72/F	Deafness	Incontinence	No	Paraganglioma/Lum	GTR	NA
15	Grunshaw 1993	29/F	Loss of taste/ smell, deafness, ataxia, myelopathy, nystagmus	No	Obstructive/ No	Unknown****/IVv	Nottreated	NA
16	Offenbacher 1996	48/M	No	No	NA	Neurinoma/FS	NA	NA
17	Castelli 1997	48/M	Deafness, ataxia	H/A, N/V	Obstructive/ No	Ependymoma/IVv	SR	NA
18	Friedman 1998	21/M	Absent	Incontinence, low back pain	No	Myx Ependymoma/ Lum	B	NA/Sphincter dysfunction, paraparesis; dissemination
19	Matsumoto 1998	48/M	Deafness, tinnitus, ataxia, myelopathy, sphincter dysfunction	No	Yes/No	Melanocytoma/T	GTR	1y/Unchanged
20	Kato 1998	24/M	Absent	Polydipsia, vertigo	No	Metastasis**/ Suprasellar	GTR + Rt	7m/Dissemination
21	Lemmerling 1998	50/M	Deafness, ataxia	No	No	Ependymoma/Lum	GTR	NA
22	Sharma 1998	60/M	Deafness, ataxia	Low back pain	No	Paraganglioma/Lum	GTR	NA
23	Durieux 1999	66/M	Deafness, ataxia	No	No	Adenoma/Sellar	SRx 2+ Rt	NA

Contd...

Table 1: Contd...

	Reference	Age/ Sex	Presentation		HCP/ Treatment	SOB/Location	Management	Follow-up/Outcome
			SS	Other				
24	Bostantjopoulou 2000	61/F	Deafness, ataxia, myelopathy	No	No	Pilocytic astrocytoma/ TL	GTR	NA
25	Straube 2001	55/F	Deafness, ataxia, polyradiculopathy, sphincter dysfunction	No	No	Pilocytic astrocytoma/ Parasellar	STR + Rs	NA
26	Das 2001	50/M	H/A, N/V, meningismus, papilledema	Myelopathy	Arreabsortive/ VPS	Melanocytoma/TL	GTR + Rt	30m/Died; Dissemination (8m)
27	Yoshida 2002	54/F	Deafness, ataxia	No	No	Teratoma/C	GTR	NA
28	Salem 2002	44/F	Deafness, ataxia	H/A, N/V, cervical pain	No	Ependymoma/IVv	GTR	NA
29	Elalaoui 2003	44/F	Deafness, ataxia	No	No	Ependymoma/IVv	STR + Rt	4m/Unchanged
30	Kitis 2003	46/M	Absent	Impaired consciousness hemianopsia	Obstructive/ No	Adenoma/Sellar	STR	NA
31	Kitis 2003	36/M	Deafness, nystagmus	Low back pain	No	Myx Ependymoma/ Lum	GTR	NA
32	Vibert 2004	55/F	Deafness, ataxia	Deep hypoestesia	No	Ependymoma/Lum	STR	4y/Progression of SS symptoms
33	Messori 2004	65/M	Deafness, ataxia, myelopathy, nystagmus	Deep hypoestesia	No	Myx Ependymoma/ Lum	GTR	2m/NA***
34	Kumar 2006	19/M	Incontinence, ataxia	Seizures, hemianopsia, hemiparesis	No	Germ cell tumor/ BBGG	B x 2 (Non Diagnostic)	6y/Died
35	Konya 2006	49/F	No	H/A	No	PGNT/FL	GTR	1y/Asymptomatic
36	Spengos 2007	63/M	Deafness, ataxia, myelopathy	No	No	Ependymoma/Lum	GTR	NA/Unchanged
37	Léveque 2009	23/M	H/A, N/V, dysphagia, dysarthria, left VI nerve palsy	Sphincter dysfunction	No	Myx Ependymoma/ Lum	GTR	3m/Improvement
38	Vreto 2011	47/M	Tinnitus	H/A, VIIcp palsy, left Romberg	No	Melanocytoma/CPA	SR	NA
39	Vyas 2011	40/M	Deafness, ataxia, dementia	No	No	Adenoma/Sellar	SR	NA
40	Steinberg 2013	43/M	No	Hemianopsia	No	Pituitary apoplexy/ Sellar	MT	NA/Decrease in size of the lesion
41	Grech 2013	64/F	Deafness, ataxia	Low back pain	No	Myx Ependymoma/ Lum	GTR + CI	NA/Improved
42	Al-Najar 2013	70/M	Ataxia	H/A, N/V, hemiparesis	No	Hemangioblastoma/LV	B + GTR	NA
43	Tosaka 2014	69/M	Deafness, ataxia, dementia	H/A, vision loss	No	Craneopharyngioma/ IIIv	STR	1y/Progression of SS symptoms
44	Pikis 2014	33/M	Deafness	Low back pain	No	Myx Ependymoma/ Lum	GTR	2y/Unchanged
45	Present case	35/M	Deafness, nystagmus, cervical pain	Drop attacks, drowsiness, ataxia, papilledema	Yes/EVD	Ependymoma/IVv	GTR	1.5y/No progression of SS symptoms

Including the present case, there are 45 published case reports of superficial siderosis secondary to nonoperated CNS tumors. 9/45 (20%) had an intraventricular location, 12/45 (27%) were located intracranially, 21/45 (47%) had a spinal location, 2 case reports did not describe the location and 1 was a carcinomatous meningitis.[1-3,6-10,12-15,19-23,26,28,30,34,36-38,41,43-45,48,50-61,63,64]
BBGG: Basal ganglia (r: right, l: left), B: Biopsy, C: Cervical spine, CI: Cochlear implant, cp: cranial pair, CPA: Cerebellopontine angle, H/A: Headache, FL: Frontal lobe (r: right, l: left), FS: Frontal sinus, GTR: Gross total resection, HCP: Hydrocephalus, Hmen: Hemangioblastic meningioma, IChA: Iron chelating agent, IIIv: Third ventricle, IVv: Fourth ventricle, LV: Lateral ventricle (r: right, l: left), Lum: Lumbar, MT: Medical treatment, NA: Not available, N/V: Nausea/vomiting, PGNT: Papillary glioneuronal tumor, Rs: Radiosurgery, Rt: Radiotherapy, Sp: Spinal, SR: Surgical resection, removal degree not specified, SS: Superficial siderosis, STR: Subtotal resection, T: Thoracic spine, TL: Thoracolumbar spine, VP: Ventricular puncture, *: Gastric carcinoma, **: Embryonal carcinoma, ***: Bilateral subdural hematomas which required surgical evacuation, ****: Suspicion diagnosis of ependymoma

all 8 cases secondary to myxopapillary ependymoma had an intraspinal location;[12-14,20,30,38,45,52] thus, it would seem that our differential diagnosis was statistically improbable. All four previously published cases of a fourth ventricular hemorrhagic mass and SS have been diagnosed as ependymoma. We suggest that, in the rare cases where this association is found, the diagnosis of ependymoma should be strongly considered. Nevertheless, the establishment of a definite radiological diagnosis represents a difficult task in such a radiological context as intraventricular cavernomas frequently lack the characteristic peripheral hypointense rim in T2-weighted MRI due to the absence of bleeding into the surrounding brain tissue.[5]

Chronic HCP has been reported in one-third of those cases presenting SS secondary to current tumors, and has been attributed to impairment of the absorption of CSF caused by subarachnoid adhesions.[39,57] Although the location of the tumor in our patient may explain by itself the development of HCP, a revision of the other 4 cases of fourth ventricle tumors with SS reveal that, in two there was no HCP,[10,50] while it was described as mild in the remaining two;[6,15] therefore, we think that a mixed etiology cannot be completely ruled out in the present case. The drop attacks experienced by our patient were probably caused by transient intratumoral bleeding, producing sudden decompensation of a preexisting HCP. The clinical tolerance of our patient to EVD removal and the presence of triventricular dilation in follow-up MRIs provide additional support for this hypothesis.

CONCLUSION

Primary or secondary SS is a rare, progressive condition that can potentially lead to severe and irreversible CNS sequelae. In the case of SS secondary to CNS tumors, the fact that its symptoms at presentation can derive from the tumor itself, SS or both, forces clinicians to be aware of the existence of this entity, since its initial manifestations may be subtle.

Current imaging technology, specifically gradient echo susceptibility T2-weighted MRI, has considerably improved our capacity to establish the diagnosis of SS. The necessity to look for a primary etiology of SS cannot be overemphasized; thus, we think it is mandatory to perform a complete examination of the CNS, once its presence has been determined, to rule out a hemorrhagic lesion. Although the optimal management remains to be determined, if a bleeding source can be established, its surgical ablation appears to halt the progression of the disease, greatly improving the prognosis of the patient.

The present article represents the fifth published case of a fourth ventricle lesion with SS. All have been found to be caused by an ependymoma. We believe that in the future cases associating a fourth ventricle lesion with secondary SS, the diagnosis of ependymoma should be strongly considered.

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Conflicts of interest
There are no conflicts of interest.

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Commentary

Re: Rare association of secondary superficial siderosis caused by a fourth ventricle hemorrhagic ependymoma mimicking a cavernoma: Case report and literature review

Secondary superficial siderosis (SS) is a rare condition in which there is iron staining of the cortical surface or ventricles depending on the location of the causative lesion. SS is not mentioned in the encyclopedic Youmans Neurological Surgery.[1] Most neurosurgeons would never have encountered a patient with SS. Neurosurgeons and neuroradiologists should be aware of it because of its potential for causing serious neurological decline.

As the authors state, SS is caused by low-grade chronic or repeated hemorrhage in the subarachnoid space. According to the authors, in 35% of the cases, the source for the hemorrhage is not identified. SS is identified on magnetic resonance imaging (MRI), as described in the article; during surgery, it appears as a brownish discoloration of the brain surface.

SS may cause a multitude of potentially serious and progressive neurological sequelae, which are well described in the article. Espinosa *et al*. have performed a thorough literature search and identified 45 cases of brain tumors associated with SS. Half of the tumors were ependymomas, and 5 of the 45 were in the fourth ventricle. Espinosa *et al*. describe an additional patient with SS caused by an ependymoma in the fourth ventricle.

The authors also point out that the ependymoma of the fourth ventricle could have similar radiological features to a cavernous hemangioma (CH) because in this location there is no typical hemosiderin ring around the CH.

The key point in management is that, when SS is identified on MRI, a thorough search for the source of hemorrhage should be undertaken and the primary pathology should be eliminated. Notwithstanding the necessity to treat the primary lesion, potentially serious neurological decline may thus be avoided.

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1. Youmans Neurological Surgery. Winn RH. 6th Ed. Philadelphia, PA: Elsevier, Saunders; 2011. ISBN 9780323249485.