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

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. The pathologic changes associated with SS have been previously described. Macroscopically, the...
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.[18]

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 tumoral 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, remodelling of the sella turcica) (c and d)
Intratumoral bleeding (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.\[^{[31]}\] 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.\[^{[11,26]}\]

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.\[^{[6,10,15,30]}\] 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.\[^{[17,18,24,25,29,30,32,35,42,47,49]}\]
| Reference       | Age/Sex | Presentation                        | HCP/Treatment | SOB/Location   | Management        | Follow-up/Outcome |
|-----------------|---------|-------------------------------------|---------------|----------------|-------------------|-------------------|
| 1 Noetzel 1940  | 47/M    | Deafness, dementia                  | No            | NA             | Metastasis*/CxMen | NA                |
| 2 Rosenthal 1958| 27/M    | NA                                  | NA            | Incontinence   | Oligodendroglioma/NA | NA                |
| 3 McGee 1962    | 54/M    | Deafness, myelopathy                | NA            | NA             | Ependymoma/Lum    | 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                |
| 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                |
| 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                |
| 12 Willeit 1992 | 59/M    | Deafness, ataxia, myelopathy, tinnitus | 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            | No             | Neurinoma/FS      | 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 1y/Unchanged |
| 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...
| Reference | Age/Sex | Presentation | HCP/ Treatment | SOB/Location | Management | Follow-up/Outcome |
|-----------|---------|--------------|----------------|--------------|------------|------------------|
| 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/ Paraspinal | STR + Rs | NA |
| 26 Das 2001 | 50/M | H/A, N/V, meningismus, papilledema | Myelopathy | Arreabsorptive/ 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, cervical pain | No | No | Ependymoma/IVv | GTR | NA |
| 29 Elalaoui 2003 | 44/F | Deafness, ataxia | No | No | Ependymoma/IVv | STR + Rs | 4m/Unchanged |
| 30 Kitis 2003 | 36/M | Absent | Impaired consciousness | Obstructive/ No | Adenoma/Sellar | STR | NA |
| 31 Kitis 2003 | 50/M | Deafness, ataxia, myelopathy | Deep hypoestesia | No | Myx Ependymoma/ Lumbar | GTR | NA |
| 32 Vibert 2004 | 55/F | Deafness, ataxia | Deep hypoestesia | No | Myx Ependymoma/ Lumbar | GTR | NA |
| 33 Messori 2004 | 65/M | Deafness, ataxia, myelopathy | Deep hypoestesia | No | Myx Ependymoma/ Lumbar | GTR | NA |
| 34 Kumar 2006 | 49/F | Incontinence, ataxia | Seizures, hemianopsia, hemiparesis | No | Germ cell tumor/ BBGG | B x 2 (Non Diagnostic) | 6y/Died |
| 35 Konya 2006 | 47/M | No | H/A | No | PGNT/FL | GTR | 1y/Asymptomatic |
| 36 Spengos 2007 | 63/M | Deafness, ataxia, myelopathy | No | No | Ependymoma/Lumbar | GTR | NA |
| 37 Léveque 2009 | 23/M | H/A, N/V, dysphagia, dysarthria, left VI nerve palsy | Sphincter dysfunction | No | Myx Ependymoma/ Lumbar | 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/ Lumbar | 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 | Craniopharyngioma/ IIIv | STR | 1y/Progression of SS symptoms |
| 44 Pikis 2014 | 33/M | Deafness | Low back pain | No | Myx Ependymoma/ Lumbar | 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 meningioma. 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), 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 of 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.[9]

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,[10,30] 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. 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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