Intramedullary Spinal Cord Hemorrhage (Hematomyelia)

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Intramedullary spinal cord hemorrhage (hematomyelia) is an uncommon cause of myelopathy and can present in an acute, subacute, stepwise, or chronic fashion. Spinal vascular malformations such as intramedullary cavernomas and intradural arteriovenous malformations are the most common cause of atraumatic intramedullary spinal cord hemorrhage based on the existing literature. Additional considerations include warfarin or heparin anticoagulation, hereditary or acquired bleeding disorders, primary spinal cord tumors, spinal cord metastases, Gowers’ intrasyringal hemorrhage, or a delayed complication of spinal radiation. Prompt diagnosis of hematomyelia first requires recognition of a myelopathy syndrome (transverse, central, anterior, posterior, or hemi-cord) often accompanied by sudden, severe back or neck pain and sometimes radicular pain. MRI with and without gadolinium is the preferred imaging modality. There are no clinical trials to guide the management of acute intramedullary spinal cord hemorrhage, and subsequent treatment is usually directed toward the underlying cause.

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Intramedullary spinal cord hemorrhage is a rare neurologic disorder that often leads to permanent disability. It has been recognized as a cause of myelopathy since the early 1800s, and the term hematomyelia was coined in 1827 to describe this condition. Although the typical presentation is one of sudden, severe, localized back pain (with or without radicular pain) and a corresponding neurologic deficit, intramedullary spinal cord hemorrhage can also present in a subacute, stepwise, or chronic fashion. Advanced imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) have facilitated the diagnosis of hematomyelia, but the underlying cause can be difficult to
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determine at the time of presentation if blood products obscure underlying pathology.

Numerous small case series and case reports describe the causes, clinical characteristics, and outcomes of patients with intramedullary spinal cord hemorrhage, but to date there has been no comprehensive literature review of hematomyelia. Thus, to better define the nature of this rare condition, we performed a review of the literature from January 1970 to December 2008 using Medline searches for the following terms: spinal cord hematoma, spinal cord haematoma, spinal cord hemorrhage, hematomyelia, haematomyelia, and spinal cord plus hemorrhage.

Causes of Hematomyelia

Traumatic Spinal Cord Injury

The most common cause of intramedullary spinal cord hemorrhage is traumatic spinal cord injury. The presence of spinal cord hemorrhage on MRI in this setting is associated with worse neurologic outcomes. Other traumatic causes of intramedullary spinal cord hemorrhage include spine surgery, minor head or spine injury, and chiropractic neck manipulation.

Spinal Vascular Malformations

The most frequently reported cause of atraumatic hematomyelia is bleeding from a spinal vascular malformation. Those vascular malformations that occur in the central nervous system include arteriovenous malformations (AVMs), cavernous angiomas (cavernomas), capillary telangiectasias, and venous angiomas. We found no cases of hematomyelia due to isolated capillary telangiectasia or venous angioma. Spinal AVMs and cavernomas, however, are well-established causes of intramedullary spinal cord hemorrhage and will each be addressed in turn.

Spinal AVMs. Spinal AVMs are classified into 4 categories (types 1, 2, 3, and 4) according to their anatomic features, location, chief arterial supply, and venous drainage. Type 1 AVMs, otherwise known as dural arteriovenous fistulas (AVFs), are thought to be acquired lesions that cause an abnormal connection between a structurally normal spinal radicular artery and an intramedullary vein. This abnormal arteriovenous connection causes dilation of the medullary venous system and venous stasis. This can lead to a congestive myelopathy and secondary cord ischemia (also known as Foix-Alajouanine syndrome), which is the most common manifestation of dural AVFs. Although rare, hematomyelia from a dural AVF can occur if there is rupture of an involved vessel or hemorrhagic transformation of a venous infarction.

AVM types 2, 3, and 4 are collectively referred to as intradural AVMs. Type 2, or glomus AVMs, are tightly compacted vascular malformations within a short segment of the spinal cord. Type 3, or juvenile AVMs, often have both intramedullary and extramedullary components with multiple feeding vessels. Type 4 AVMs, or intradural perimedullary AVFs, are located on the surface of the spinal cord. These intradural AVMs are thought to be congenital malformations that are comprised of structurally abnormal vessels. Without the benefit of an interposing capillary bed, the abnormally thin vessels in these malformations can form aneurysmal dilatations or rupture when exposed to high-pressure arterial blood flow, causing spinal subarachnoid hemorrhage and, less often, hematomyelia. Hemorrhagic transformation of a venous infarct may also contribute to hematomyelia from intradural AVMs, as pial venous reflux and venous congestion are frequently observed in this setting.

The risk of hemorrhage from spinal intradural AVMs has not been well studied. In a retrospective series of 32 patients with intramedullary AVMs, 7 of the 10 children (including 5 with hereditary hemorrhagic telangiectasia) and 6 of the 22 adults presented with intramedullary spinal cord hemorrhage (70% and 27%, respectively), whereas 45% to 48% of patients with intracranial AVMs presented with hemorrhage.

Spinal cavernomas. Spinal cavernomas are infrequent compared with their intracranial counterparts. Hemorrhage from an intradural cavernoma typically results in sudden neurologic decline, although recurrent microhemorrhages can also present as a chronic progressive myelopathy. Hematomyelia is most often caused by intramedullary cavernomas, but can also be caused by extramedullary intradural cavernomas.

Cavernomas have a typical appearance on MRI (a well-circumscribed popcorn-like lesion with a heterogeneous core surrounded by a dark rim of hemosiderin on T1-weighted images), but this may be obscured by blood products at the time of initial imaging, making diagnosis difficult (Figure 1). Because of slow blood flow through these lesions, cavernomas are typically not visible on formal angiograms. They are thus referred to as angiographically occult.

Only about 5% of patients with an intracranial cavernoma will also have a spinal cavernoma. Conversely, 45% of patients with a spinal cavernoma will also have an intracranial cavernoma. Only very rare patients have multiple spinal cavernomas without any accompanying intracranial cavernomas. Thus, if a spinal cavernoma is suspected or identified, an
additional MRI of the head should be considered.

In large retrospective reviews of patients with symptomatic spinal cord cavernomas, 36% to 58% present with spinal cord hemorrhage.\(^1,30\) The annual risk of hemorrhage from intramedullary spinal cord cavernomas is estimated to be 1.4% per lesion per year (using the age of symptom onset and assuming that all cavernomas were present at birth) according to a retrospective review of 111 cases.\(^1\) A more recent retrospective review of 33 patients found the rate of symptomatic hemorrhage to be 1.6% per lesion per year.\(^27\) These estimates are similar to the risk of bleeding from intracranial cavernomas, which ranges from 0.7% to 2.5% per lesion per year.\(^31-33\) Spine trauma, chiropractor manipulation, or exertion can seemingly precipitate hemorrhage from an intramedullary cavernoma in rare cases.\(^34\)

**Anticoagulation**

Intracranial hemorrhage is a feared complication of anticoagulation therapy. Although much less common, hematomyelia has the potential to be equally devastating. Anticoagulation-related intramedullary spinal cord hemorrhage may be the first presentation of an occult cord lesion,\(^35\) or it may occur at the site of cord inflammation (ie, at the level of the spinal cord corresponding to a cutaneous eruption of herpes zoster).\(^36\) Most reported cases, however, are not associated with any underlying pathology or precipitating trauma.\(^37-45\) Although the degree of anticoagulation is correlated with the risk of hemorrhagic complications, warfarin-associated hematomyelia can occur when the prothrombin time (PT) or international normalized ratio (INR) is within or very near the therapeutic range (the therapeutic PT is considered to be 25 ± 5 seconds, and the therapeutic INR is considered to be 2-3).\(^37,38\)

Apart from warfarin anticoagulation, there is 1 reported case of hematomyelia after attempted spinal anesthesia in a patient who had received low-dose subcutaneous heparin 1 hour earlier.\(^44\)

**Hereditary or Acquired Bleeding Disorders**

Like pharmacologic anticoagulation, hereditary or acquired bleeding disorders can also cause intramedullary spinal cord hemorrhage. The most common hereditary bleeding disorders include von Willebrand disease and X-linked factor VIII deficiency (hemophilia A). Acquired bleeding disorders typically occur in the setting of chronic liver disease, vitamin K deficiency, malignancy, autoimmune disease, or clotting factor inhibitors.

Bleeding disorders associated with hematomyelia documented in the literature include von Willebrand disease,\(^45\) factor VII deficiency,\(^46\) a factor VIII inhibitor,\(^47\) and factor XI deficiency.\(^48\)

**Primary Spinal Cord Tumors and Metastases**

Primary intramedullary spinal cord tumors typically present with gradually progressive neurologic signs and symptoms that vary depending on the location of the lesion; only rarely do they present with hematomyelia. Those primary spinal cord tumors reported to cause intramedullary spinal cord hemorrhage include ependymoma,\(^35,49,50\) fibrillary astrocytoma,\(^51\) and primary central nervous system melanoma.\(^52\) Hemorrhage into an ependymoma of the filum terminale has also presented as a cauda equina syndrome.\(^53\)

Hemorrhagic spinal cord metastases can also cause hematomyelia. Spinal cord metastases are rare, accounting for 4% to 8.5% of central nervous system metastases\(^54\) and causing only 6% of myelopathies in cancer patients.\(^55\) Lung cancer accounts for half of all spinal cord metastases, followed by breast cancer in 12.5%.\(^55\) Although renal cell carcinoma accounts for only 5% of all spinal cord metastases,\(^55\) to our knowledge it is the only type of cord metastasis reported to present with intramedullary spinal cord hemorrhage.\(^54,56\)

Approximately 50% to 60% of patients with a metastatic intramedullary spinal cord lesion also have metastatic brain lesions.\(^55\) Thus, identification of a spinal cord metastasis should prompt imaging of the brain as well.

**Gowers Intrasyringal Hemorrhage**

The first case of hematomyelia caused by spontaneous hemorrhage...
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<table>
<thead>
<tr>
<th>Cause</th>
<th>Historical Clues</th>
<th>MRI Features</th>
<th>Specific Treatments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spine trauma</td>
<td>Motor vehicle accident, Fall or other trauma, Diving or sports injury</td>
<td>Associated trauma to other spinal structures</td>
<td>Spinal cord decompression and spine stabilization, Steroids (debated)</td>
</tr>
<tr>
<td>Spinal arteriovenous malformation</td>
<td>Sudden-onset neurologic deficits, Severe headache or back pain with meningismus if associated subarachnoid hemorrhage</td>
<td>Abnormal tortuous vessels or flow voids</td>
<td>Endovascular embolization, Surgical resection</td>
</tr>
<tr>
<td>Spinal cavernoma</td>
<td>Personal or family history of cavernomas</td>
<td>Popcorn-like lesion of variable intensities surrounded by a dark hemosiderin ring</td>
<td>Observation, Surgical resection</td>
</tr>
<tr>
<td>Anticoagulation</td>
<td>Use of warfarin or heparin, Elevated INR or aPTT, Sudden onset of pain or neurologic deficit</td>
<td>Possible air-fluid levels</td>
<td>Reversal of anticoagulation</td>
</tr>
<tr>
<td>Genetic bleeding diathesis</td>
<td>Personal or family history of excess bleeding, Male gender (in the case of hemophilia)</td>
<td></td>
<td>Administration of fresh frozen plasma or specific clotting factors</td>
</tr>
<tr>
<td>Acquired bleeding diathesis</td>
<td>Liver disease, Known malignancy or autoimmune disease, Postpartum period</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary spinal cord tumor</td>
<td>Progressive neurologic deficit</td>
<td>Mass effect with cord enlargement, Abnormal gadolinium enhancement</td>
<td>Surgical resection, Radiation</td>
</tr>
<tr>
<td>Spinal cord metastasis</td>
<td>Known systemic cancer, especially renal cell carcinoma</td>
<td>Mass effect with cord enlargement, Gadolinium enhancement</td>
<td>Surgical resection, Radiation</td>
</tr>
<tr>
<td>Intrasyringal hemorrhage</td>
<td>Pre-existing syringomyelia, Condition known to predispose to syrinx formation (eg, Arnold-Chiari malformation, past meningitis, trauma, scoliosis, etc)</td>
<td>Cervicothoracic location, Low-lying cerebellar tonsils if associated Arnold-Chiari malformation</td>
<td>Possible surgical decompression</td>
</tr>
</tbody>
</table>

MRI, magnetic resonance imaging; INR, international normalized ratio; aPTT, activated partial thromboplastin time.

into a pre-existing spinal cord syrinx was described by Gowers in 1904 and has been infrequently reported since. It is theorized that a sudden increase of pressure within the syringomyelic cavity may cause vessels that are traversing the syrinx to rupture and bleed. Spontaneous intrasyringal hemorrhage can cause acute, episodic, or gradually progressive neurologic decline and should be considered in patients with conditions that predispose to syrinx formation (eg, Arnold-Chiari malformation, scoliosis, arachnoiditis, past
trauma or meningitis). Mild trauma can also precipitate hemorrhage into a pre-existing syringomyelia.

Other Rare Causes
Other rare causes of hematomyelia in the literature include hemorrhagic infarction associated with abdominal aortic aneurysm or fibrocartilaginous emboli, sudden intracranial hypertension due to massive intracerebral and intraventricular hemorrhage, aortic coarctation, following streptokinase treatment of myocardial infarction, as a delayed complication of spinal radiation, and in patients with congenital atlanto-axial dislocation at the moment of or just after surgical decompression of the high cervical spinal cord. Herpes simplex virus myelitis is a theoretical cause of hematomyelia given its propensity to cause hemorrhagic necrosis, as is primary central nervous system vasculitis involving the spinal cord—although no cases of hematomyelia associated with these conditions were identified in the literature. Likewise, no adult cases of hematomyelia related to thrombocytopenia were identified.

Diagnosis
Heightened clinical suspicion for hematomyelia is required due to its rarity and varied presentations. Prompt diagnosis of hematomyelia first requires recognition of a myelopathy syndrome (transverse, central, anterior, posterior, or hemi-cord) often accompanied by sudden, severe back or neck pain—with or without radicular pain. A careful history can provide additional clues to the diagnosis and identify potential risk factors for intramedullary spinal cord hemorrhage (Table 1). The next step in the evaluation of myelopathy—especially if it presents in a sudden or rapidly progressive fashion—is to obtain an imaging study. At present, MRI with and without gadolinium is the preferred imaging modality because it can identify not only hematomyelia but also other causes of myelopathy—including compressive mass lesions, which are important to consider in the differential diagnosis of any sudden or rapidly progressive myelopathy. Newer MRI techniques, including gradient-echo and susceptibility-weighted sequences, are more sensitive than conventional T1- and T2-weighted images in detecting acute intracranial hemorrhage and may have applications in spinal imaging as well. If hematomyelia is present, MRI is also often able to provide clues to the underlying etiology of the hemorrhage (Table 1). Spinal CT is another alternative if MRI imaging is not available or is contraindicated. Finally, initial laboratory studies should include a complete blood count, INR, and activated partial thromboplastin time.

Mechanism of Injury
Intramedullary spinal cord hemorrhage creates a mass lesion in the substance of the cord. Because of the anatomic structure of the spinal cord, blood tends to dissect longitudinally above and below the area of initial hemorrhage, disrupting gray matter more than white matter and sometimes extruding out into the nerve rootlets (Figure 2). A complex cascade of events then occurs. Mechanical disruption of cord tissue compresses adjacent cord structures, and pressure from this compression decreases blood flow to surrounding cord tissue. Breakdown of the blood-spinal cord barrier causes vasogenic edema. Release of inflammatory mediators impairs vascular autoregulation and can cause vasospasm. The cumulative effect of these various pathologic processes is what ultimately results in spinal cord injury.

Figure 2. Gross intraoperative photograph demonstrating severe hemorrhage in the conus medullaris with blood extruding out of multiple enlarged dorsal rootlets.

This tissue response to the injury after hematomyelia occurs explains why the neurologic deficits in some patients continue to worsen for hours or even days after the initial bleed. The radiologic correlate of this process can manifest as T2 hyperintensity within the substance of the cord, cord swelling, and gadolinium enhancement (Figure 3).
Management

There are no clinical trials to guide the acute management of hematomyelia to minimize cord injury. Many advocate hematoma evacuation, but which patients stand to benefit most from surgery and the optimal timing of clot evacuation are unknown.\textsuperscript{41,63,77} For example, some argue that surgery should be done immediately to remove mass effect and pressure on the spinal cord, whereas others prefer to let any neurologic deficits plateau before removing the clot to avoid damaging viable surrounding tissue. There is at least 1 case report of a patient improving after high-dose methylprednisolone,\textsuperscript{18} but there is not enough evidence to argue for or against this intervention.

Subsequent treatment is usually directed toward the underlying cause of the hematomyelia (Table 1). Examples include endovascular embolization of spinal AVMs,\textsuperscript{17,20} surgical resection of symptomatic spinal cavernomas,\textsuperscript{24,28,30} reversal of anticoagulation,\textsuperscript{19} or correction of a hereditary or acquired bleeding diathesis.\textsuperscript{46,47}

As in spinal cord compression, prognosis in hematomyelia is thought to correlate most with the patient's neurologic status at the time of presentation.

Conclusion

Based on the existing medical literature, the most common cause of atraumatic intramedullary spinal cord hemorrhage is bleeding from a spinal vascular malformation, such as intradural AVMs (types 2, 3, and 4) or intramedullary cavernomas. Additional

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Main Points

- Intramedullary spinal cord hemorrhage (hematomyelia) can present as an acute, subacute, stepwise, or chronic myelopathy.
- Traumatic spinal cord injury is the most common cause of hematomyelia.
- Atraumatic hematomyelia is rare and often caused by spinal vascular malformations, including intradural arteriovenous malformations or cavernomas.
- Anticoagulation, hereditary or acquired bleeding disorders, primary or metastatic spinal cord tumors, and bleeding into a pre-existing syrinx are additional considerations.
- Magnetic resonance imaging with and without gadolinium is the preferred imaging modality, although blood products can obscure underlying pathology.
- Neurologic deterioration can occur after the initial hemorrhage due to the secondary tissue response to injury.
- Treatment is usually directed toward the underlying cause.
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considerations include warfarin or heparin anticoagulation (even in the absence of an underlying cord lesion or precipitating trauma), hereditary or acquired bleeding disorders, primary spinal cord tumors or spinal cord metastases (specifically renal cell carcinoma, which is known to be a highly vascular tumor), bleeding into a pre-existing spinal cord syrinx (also known as Gowers intrasyringal hematomyelia), or a delayed complication of spinal radiation.

Although the clinical presentation often is one of sudden neurologic decline, intramedullary spinal cord hemorrhage can present in a variety of ways and should thus be considered in the differential diagnosis of acute, subacute, stepwise, or chronic myelopathy. There are no clinical triads to guide the management of acute intramedullary spinal cord hematomyelia, and subsequent treatment is usually directed toward the underlying cause.

References


