**FORENSIC SCIENCE NEWSLETTER**
**Forensic Pathology and Neuropathology**

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## FORENSIC PATHOLOGY

### Dieulafoy's Lesion: An Uncommon Cause of Sudden Massive Gastrointestinal Hemorrhage

The Dieulafoy lesion is an unusually large submucosal artery typically found in the proximal portion of the stomach, along the lesser curvature (80-85%), usually within 6 cm of the esophagealgastric junction. They can also be found in the rectum and colon (10%), small bowel (2%), and the esophagus (2%). Dieulafoy's lesion can manifest clinically as recurrent gastrointestinal hemorrhage or a sudden, massive gastrointestinal hemorrhage, which on occasion can lead to death. This lesion makes up to 1-2% of gastrointestinal bleedings.

Dieulafoy's lesion, also referred to as a **cirsoid aneurysm, submucosal arterial malformation** and initially as **exulceratio simplex**. It was originally described by Gallard in 1884 as a "miliary aneurysm" of the stomach. George Dieulafoy, a French Surgeon, identified the same lesion in 1898 in three patients with massive upper gastrointestinal hemorrhage. Originally, Dieulafoy referred to the lesion as "exulceratio simplex," which was later changed to Dieulafoy's lesion.

As Gallard described the lesion, it was originally thought to represent an aneurysm in the gastric submucosal arteries, perhaps in combination with atherosclerosis. It was then suggested that it represented either a congenital or acquired vascular malformation. Today, most believe it is an abnormally large-caliber vessel, ranging from 1-5 mm in diameter, most consistent with a tortuous submucosal artery. It has been speculated that the presence of a large superficial submucosal arteriole causes continuous pressure on the overlying mucosa, leading to its thinning and rendering it more susceptible to traumatic ulceration and vessel erosion with time. This would explain its usual appearance in adults and its association with either recurrent gastrointestinal hemorrhage or massive hemorrhage, which can be lethal.

There are other theories as to the genesis of spontaneous bleeding. One suggest the continued pulsation of the submucosal venous vessels causes damage to the overlying epithelium which in turn leads to focal ischemia. This is followed by erosion and vascular rupture. Another theory is the abnormally large-caliber arteriole develops a thrombosis, which in turn causes focal necrosis followed by rupture and bleeding. It has also been suggested for those large superficial arterioles, which occur in the small and

large bowel and rectum, the solid content within their lumens leads to the development of mucosal ulceration and eventual erosion of the vessel wall. Since, these lesions occur more commonly in the older age group, some believe the mucosal atrophy associated with advancing age may contribute to vessel wall erosion.

When these lesions are examined with the microscope they show abnormally large-caliber arterioles with evidence of aneurysm formation, atherosclerosis, and inflammation in the vessel wall.

Clinically, these patients present with recurrent hematemesis (vomiting of blood with the source generally in the upper gastrointestinal tract) with melena (tarry feces [black color] due to the hemoglobin from the upper gastrointestinal bleeding being altered by intestinal chemicals and bacteria), which is present in 51%; hematemesis without melena present in 28%; and melena alone in 18%. Those patients whose lesion are in the small bowel, colon or rectum, typically, present with hematochezia (passage of fresh bright red blood through the anus, usually in or with stools) in 3% of cases.

Typically, these lesions are seen in the older age group, with a male-to-female ratio of 2:1, who have multiple comorbidities, such as cardiovascular disease, hypertension, chronic kidney disease, and diabetes. However, it can also be seen in children, the first case of which was reported in 1968 by Rossi *et al.,* in a 20-month-old girl with Christmas disease (Hemophilia B due to mutation of factor IX gene, leading to a deficiency of factor IX). In 1986, Veldhuyzen *et al.,* published the first review of 101 patients with the Dieulafoy lesion, two of which were children. The lesions occur more frequently in boys (male: female ratio of 1.5:1) presenting at an age starting from birth, the youngest of which were two neonates, 19 and 22 hours old. This supports the concept these lesions are congenital superficial submucosal large-caliber arterioles that erode easily on ulceration of the overlying mucosa.

The diagnosis is typically made through endoscopy, with 49% of the lesions being identified on the first endoscopic procedure. 33% require more than one endoscopic procedure. If the endoscopic procedure is not successful in identifying the Dieulafoy lesion than angiography is often used for the diagnosis, especially for lesions in the colon and rectum.

Typically, these lesions are treated endoscopically, either using thermal electrocoagulation, local epinephrine injection and sclerotherapy, and clip application. Should these methods prove unsuccessful, then surgical resection might be necessary.

**Neuropathology**

**Invasive Intracranial Pressure Monitoring**

Patients with traumatic brain injury (TBI), hydrocephalus, intracranial tumors, hepatic encephalopathy, and cerebral edema often have elevated intracranial pressure (ICP) for which monitoring of the ICP becomes necessary. Intractable elevated ICP can lead to death or devastating neurological damage either by reducing cerebral perfusion

pressure causing cerebral ischemia or by compression of important brain structures leading to transtentorial herniation of the brainstem or transfalcine herniaiton.

Transtentorial herniation can cause injury to the outer fibers of the ipsilateral oculomotor nerve due to strangulation of the nerve between herniating tissue and the medial petroclinoid ligament; stretching of the nerve over the clivus from the lateral displacement of the midbrain; entrapment of the nerve between the posterior cerebral and superior cerebellar arteries from the downward displacement of the midbrain; creasing of the contralateral cerebral peduncle (Kernohan's notch or more properly, the Kernohan-Woltman phenomenon), causing a Babinski sign ipsilateral to the hemispheral lesion due pressure of the laterally displaced midbrain against the sharp edge of the tentorium; lateral flattening of the midbrain and zones of necrosis; secondary hemorrhages in the tegmentum and base of the subthalamus, midbrain, and upper pons (Duret hemorrhages); unilateral or bilateral infarction (hemorrhagic) of the occipital lobes due to compression of the posterior cerebral artery against the tentorium by the herniating temporal lobe; and rising intracranial pressure (ICP) and hydrocephalus due to lateral flattening of the aqueduct and third ventricle and blockage of the perimesencephalic subarachnoid space.

Transfalcine herniation (Cingulate herniation) is the most common type of herniation and causes the innermost part of the frontal lobe to be scraped under part of the falx cerebri. Such herniation may interfere with the blood vessels in the frontal lobes, the anterior cerebral artery resulting in a substantive increase in ICP that can lead to more significant forms of herniation, such as central herniation. Symptoms for transfalcine herniation are not as clearly defined, however, they may present as abnormal posturing (decorticate, decerebrate and opisthotonus) and coma.

Recognition of elevated ICP is important to prevent morbidity or mortality.

There are two primary techniques for measuring ICP: invasive and noninvasive. The noninvasive methods include transcranial Doppler, tympanic membrane displacement, optic nerve sheath diameter, CT scan/MRI and fundoscopy. The invasive methods include ventriculostomy (extracranial [external] ventricular drains) [EVDs] and microtransducers (intraparenchymal ICP monitors). Ventriculostomy is considered the gold standard with the most accurate measurement of pressure, although microtransducers are typically just as accurate. The invasive techniques are associated with risk of complications, such as hemorrhage and infection. Although, the noninvasive techniques are without the risk of the invasive techniques complications, they are not as accurate in measuring ICP.

In this **Newsletter** we will discuss the **microtransducer (intraparenchymal) ICP monitor.**

Typically, invasive intraparenchymal ICP monitor is inserted in patients with severe traumatic brain injury with a Glasgow Coma Scale score (GCS) between 3-8 who show abnormalities on CT scans, such as hematomas, contusions, swelling, herniation, or

compressed basal cisterns. Invasive monitoring is also done in those patients with a GCS of 3-8, but have a normal CT scan, who have two or more of the following conditions: older than 40-years-of-age, have clinical manifestations of either unilateral or bilateral abnormal motor posturing (decorticate, decerebrate and opisthotonus) or hypotension (systolic blood pressure less than 90 mmHg).

Invasive intracranial pressure (ICP) monitoring uses a device (microtransducer), placed inside the head. The microtransducer senses the pressure inside the skull and sends measurements to a recording device. Typically, the ICP microtransducer is placed in the right frontal region at a depth of approximately 2 cm. However, microtransducers can be placed within the ventricles, epidural, subdural or subarachnoid compartments.

As pointed out above, microtransducers are just as accurate as EVDs, however, microtransducers generally cannot undergo recalibration after placement. The exception to this is the Spiegelberg catheter, which recalibrates itself every hour. The EVDs, however, can be reclibrated at any time, by resetting the transducer to atmospheric pressure at the level of the so called zero reference point (Foramen of Monro/Targus). Another point to consider when comparing ICP microtransducers and EVDs, is although microtransducers monitors do not allow for therapeutic drainage of cerebrospinal fluid (CSF), the placement is less technically challenging, especially in patients with TBI, who have ventricular displacement, various degrees of collapse or effacement.

ICP monitoring, as is true of EVDs should not be used in patients with a coagulopathy (i.e., those patients with a platelet count less than 100,000 per $mm^3$, platelet dysfunction, or an international normalized ratio greater than 1.3). International normalized ratio (INR) is a standardized number that is computed in a laboratory. Knowing a patient's INR is especially important if they are taking anti-clotting medicines (blood thinners). The INR is computed from the results of the prothrombin time (PT), which measures the time it takes for your blood to clot. The INR is an international standard for the PT. Also, ICP monitor should not be inserted at or near a site of local infection.

Insertion of the ICP monitors are typically done on the side of the non-dominant hemisphere, which is usually the right side. The insertion is made 3 cm lateral to the midline (i.e., midpupillary line) to avoid the sagittal sinus; approximately 11 cm posterior to the nasion (intersection of the frontal bone and the two nasal bones of the skull, just inferior to the glabella); and at least 1 cm anterior to the coronal suture to avoid the motor strip of the underlying cerebral hemisphere and to a depth of 2 cm in the right frontal lobe.

The motor strip refers to the primary motor cortex (Brodmann's area 4), which in humans is located in the dorsal portion of the frontal lobe. It works with other motor areas of the brain including the premotor cortex, the supplementary motor area, the posterior parietal cortex, and several subcortical regions, to plan and execute movements. Within the primary motor cortex, at the microscopic level, are large

neurons called Betz cells. Betz cells, along with other cortical neurons, send axons down to the spinal cord to synapse (make a connection) onto the interneuron circuitry of of the spinal cord and directly onto the alpha motor neurons in the spinal cord, which connect to the muscles.

There are three potential complications: Hemorrhage within the scalp or skull, which usually ceases once the monitor is in place; intracranial hemorrhage is an uncommon complication (catheter-related hemorrhages occur in 1-33% of patients) and if suspected, an unenhanced CT of the head should be performed; and infection which is rare (occurs in 1-12% of patients), but should it occur, it does so late and is regarded as a serious complication. If it is suspected there is an intracranial abscess and or ventriculitis, a CSF sample should be taken as well as a CT of the head, with or without contrasts.

The normal ICP varies with age and body posture, but is typically between 5-15 mmHg in a healthy supine adult, 3-7 mmHg in children and 1.5-6 mmHg in infants. In adults, the upper limit of the normal range is 20 mmHg.
In infants, the ICP is normally maintained at a level that is very low by standards that apply later in life. There is little or no overlap of normal pressure and the pressure in infantile hydrocephalus. Remember, during the first few days after birth, as the volume of the brain decreases so does its turgor, and subatmospheric ICP is common. Hydrocephalus may be masked or attenuated in severity during that time or may be incorrectly suspected in a normal child because of the increase in circumference of the head accompanying the restitution of volume.

With an ICP monitor in place, gentile pressure over the jugular veins should cause a rise in intracranial pressure. If there is no increase in intracranial pressure, check the waveform. If an ICP waveform is not present, the ICP sensor and transducer must be checked. Technical malfunction or loss of calibration of the ICP sensor can lead to inaccurate readings. What is most important, should there be a loss of waveform, loss of calibration or any technical malfunction in which the ICP is no longer consistent with the clinical picture or radiologic findings of the patient, then the ICP sensor should be replaced. Failure to do so may lead to an elevated ICP resulting in herniation leading to irreversible brain damage and death. Treatment designed to lower ICP should be initiated at pressures above 15-20 mmHg, depending on the cause of the elevated pressure.