15 Neurointervention in Ophthalmologic Disorders

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Cerebral Aneurysm

Introduction

Cerebral aneurysms are localized abnormal dilatation of the cerebral artery resulting from weakening of the vascular wall. Cerebral aneurysms most commonly involve the branch points of the major arteries at the circle of Willis [1]. The overall prevalence of intracranial aneurysms varies from 1% to 5% [1–3]. The majority of small cerebral aneurysms (less than 7 mm in diameter) remain unruptured, with an average annual rate of rupture of approximately 0.95%; patient age, smoking, and size of aneurysm are important risk factors for rupture [4, 5]. The annual rate of rupture increases to 3–8% for aneurysms greater than 7 mm in diameter [6].

Clinical Manifestations

The ocular manifestations of cerebral aneurysms take three forms: [1] visual loss as a result of retinal/vitreous hemorrhage associated with subarachnoid hemorrhage, also called Terson syndrome, [2] visual field defects caused by aneurysmal compression of the anterior visual pathways, and [3] diplopia and ocular motility deficits caused by aneurysmal compression on the cranial nerves [7–9]. Other rare ocular manifestations have been reported, as a result of either the cerebral aneurysm or its related intervention, such as retinal arterial emboli [10], orbital compartment syndrome and blindness [11], and internuclear ophthalmoplegia [12].

Terson syndrome manifests as multilayer bleeding in the subretinal space, in the retina, between retina and vitreous, and in the vitreous and occurs in more than 30% of patients who survive subarachnoid hemorrhage [13]. The mechanism of intraocular bleeding is postulated to be rupture of the fine optic nerve head and retinal capillaries when central retinal venous stasis develops as a result of suddenly elevated intracranial pressure from subarachnoid hemorrhage. Terson syndrome commonly affects both eyes, and severity of the visual loss depends on the extent of the bleeding and its relative location to the retina (Fig. 15.1). The visual outcome in Terson syndrome is usually favorable; complete visual recovery occurs within 12 months once the blood is reabsorbed. However, the presence of Terson syndrome may predict poorer neurologic outcome from subarachnoid hemorrhage [14].

The intimate relationship between the anterior visual pathways and the circle of Willis makes visual loss a not infrequent presenting symptom of cerebral aneurysms. Cerebral aneurysms aris-

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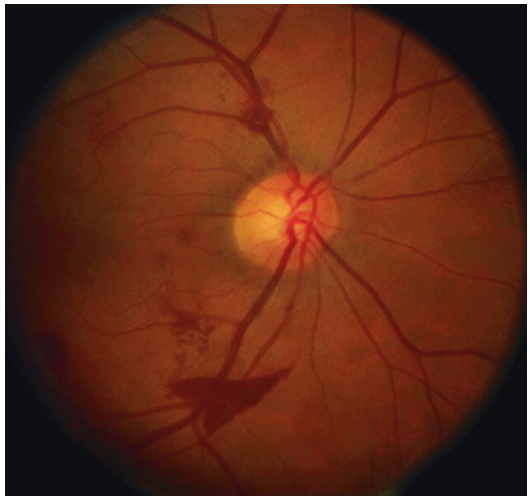

Fig. 15.1 Retinal and subhyaloid (between vitreous face and retina) hemorrhage in a patient with Terson syndrome as result of a ruptured middle cerebral artery aneurysm. Note typical appearance of triangular-shaped or boat-shaped subhyaloid hemorrhage in inferior retina

ing from intracranial carotid artery, middle cerebral artery, and anterior communicating arteries are more likely associated with visual loss than aneurysms arising from basilar and posterior communicating aneurysms [8]. Involvement of the optic nerve gives rise to decreased central vision and optic atrophy. Compression of the optic chiasm results in bitemporal field defects. Aneurysms growing posteriorly may cause homonymous hemianopia from involvement of the optic tract. Ophthalmic and supraclinoid segment carotid aneurysms constitute less than 10% of all cerebral aneurysms [5, 15]. They may present with headache, orbital pain, and visual loss by compression and without rupture. Middle cerebral artery and anterior communicating artery aneurysms account for nearly half of all cerebral aneurysms. They tend to rupture and present with subarachnoid hemorrhage before enlarging sufficiently to affect visual function [16].

Diplopia and ocular motility abnormalities are commonly associated with aneurysms arising from cavernous carotid segment, posterior communicating, and basilar arteries [17]. Posterior communicating artery aneurysm is a frequent cause of third nerve palsy, which presents with a symptom complex of ptosis, fixed and dilated pupil, and the so-called down and out eye [18]. The pupil reactivity to light stimulus should be meticulously examined to help differentiate aneurysmal compression from an ischemic third nerve palsy. A complete third nerve palsy that spares the pupil is due to an ischemic third nerve palsy, while pupil involvement in combination with either a complete or an incomplete third nerve palsy raises a high suspicion for aneurysmal compression. An incomplete third nerve palsy without pupil involvement warrants close observation over the following 1 week for development of pupil abnormalities. Aneurysms within the cavernous sinus can affect the multiple cranial nerves that travel through the cavernous sinus; the ocular motor abnormalities are sometimes associated with decreased sensation or pain along the V1 and V2 distributions of the trigeminal nerve [9]. Cavernous segment carotid aneurysms may occasionally present with a sixth nerve palsy in combination with Horner syndrome, localizing the lesion to the cavernous sinus. Basilar aneurysms, while less common, constitute the majority of cerebral aneurysms arising within the posterior fossa. Basilar artery aneurysms may cause diplopia via third, fourth, and sixth nerve involvement, skew deviation, and gaze palsy secondary to aneurysmal compression of the midbrain and pons. Basilar artery aneurysms can also cause homonymous visual field loss due to thromboembolic infarction to the occipital lobe [17].

The radiographic features and endovascular intervention for cerebral aneurysm are described in Chaps. 10 and 11.

Carotid-Cavernous Fistula

Introduction

The cavernous sinuses are a pair of cerebral venous sinuses located at the center of the skull base lateral to each side of the sella turcica (Fig. 15.2). They are bordered by the sphenoid and temporal bones. The cavernous sinus collects venous blood drained from the eye and orbit through the superior and inferior ophthalmic

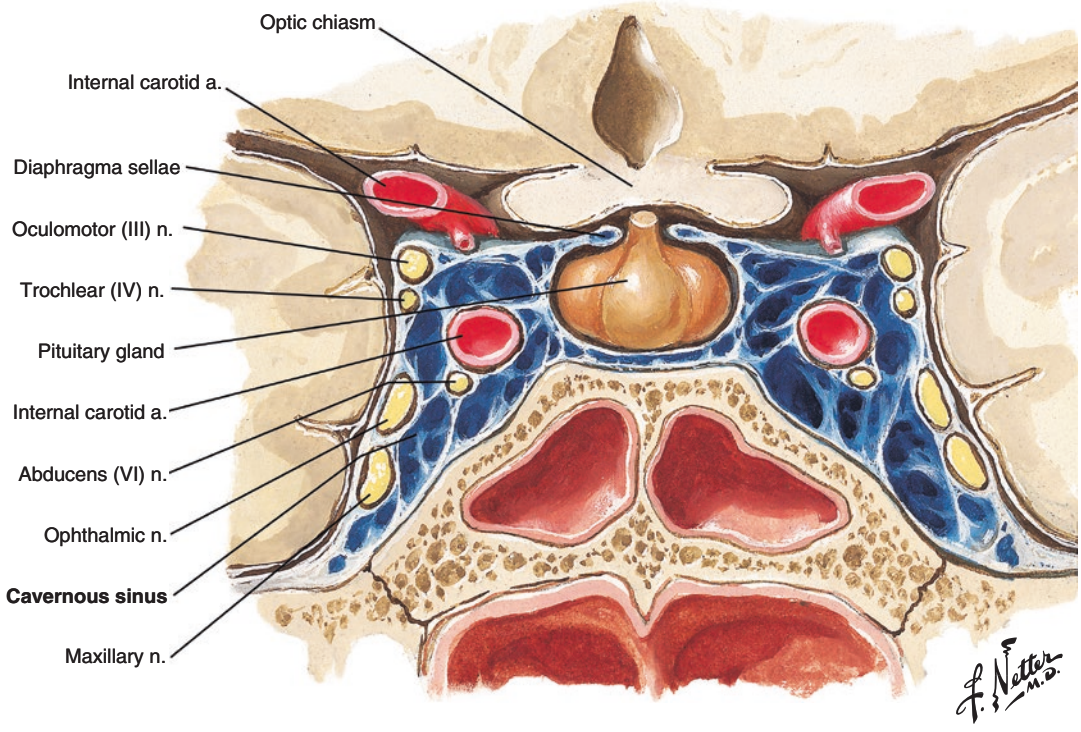

Fig. 15.2 Coronal view of the cavernous sinus demonstrating the passage of the third, fourth, fifth, and sixth cranial nerves and internal carotid artery inside the cavernous sinuses. (Permission from Netter's production)

veins and then drains posteriorly to the internal jugular vein through the superior and inferior petrosal sinuses and the transverse sinus [19]. A number of cranial nerves and intracranial vessels travel through the cavernous sinus. The cavernous segment of the internal carotid artery is located in the medial aspect of the cavernous sinus and is surrounded by oculosympathetic fibers that form a fine plexus. Immediately lateral to the internal carotid artery is the sixth cranial nerve. The third and fourth cranial nerves and the first and second divisions of the trigeminal nerve (ophthalmic and maxillary nerve, respectively) travel along the lateral border of the cavernous sinus. The pituitary gland is located in the sella turcica between the pair of cavernous sinuses.

Carotid-cavernous fistula (CCF) is an abnormal communication between the cavernous venous sinus and the carotid arterial system. Meningeal branches arising from the internal carotid artery, external carotid artery, or both supply the dural sheath and anatomical structures contained in the cavernous sinus. When there is a breach in either the main trunk of the cavernous segment of the internal carotid artery or the meningeal branches from the internal or external carotid artery, an abnormal communication develops between the arterial and venous circulation [20]. Communication between the arterial and venous system results in elevated venous pressure thus elevated venous outflow resistance. Functional obstruction of the venous drainage from the eye and orbit ensues. The classification of CCF is based on anatomy (direct vs. indirect), cause (traumatic vs. spontaneous), or hemodynamic status (high flow vs. low flow). Each type of CCF is associated with specific clinical manifestations, treatment strategies, and outcomes. The most commonly used dichotomies in CCF classification are (1) *direct CCF* formed by direct connection between the cavernous segment of the internal carotid artery and cavernous sinus and (2) *indirect CCF* caused by a communication between the branches of the internal or external

carotid arteries and the cavernous sinus. Barrow et al. [21] defined four types (Types A–D) of CCFs. Type A CCFs are direct, high-flow lesions connecting the ICA directly to the cavernous sinus. Type A CCFs often result from a single tear in the carotid artery wall, caused either by trauma or aneurysm rupture. Type B, C, and D CCFs are all indirect, low-flow lesions that arise from meningeal branches of either the ICA or ECA. Another angiography-based classification system is proposed for dural AV fistulas – the Cognard classification based on the direction of dural sinus drainage (antegrade or retrograde), the presence or absence of cortical venous drainage, and venous outflow architecture [22].

Clinical Manifestation

A direct CCF is caused by a tear in the intracavernous segment of the internal carotid artery, usually in the setting of trauma, although in a small proportion of patients, it may occur spontaneously [23]. The classic triad of proptosis, conjunctival chemosis, and orbital bruit is a consequence of significantly elevated venous pressure in the superior ophthalmic vein and cavernous sinus system [24]. Typical ocular findings are prominent pulsatile proptosis, chemosis, hyperemia, and periorbital pain or headache [25, 26]. Elevated intraocular pressure and secondary glaucoma are caused by increased episcleral pressure and vortex venous pressure, anterior shift of the lens-iris diaphragm, as well as neovascular glaucoma secondary to ocular ischemia. Ophthalmoplegia can be a consequence of either edema of the extraocular muscles or damage to the cranial nerves as they travel through the cavernous sinus. Vision loss is common and is usually severe in direct CCF, caused by exposure keratopathy, glaucoma, ischemia of the optic nerve, or coexisting traumatic optic neuropathy [27–29].

In contrast to direct CCF, indirect CCFs usually have a spontaneous onset and are slowly progressive [30]. A history of minor trauma is reported in a small group of patients. When compared to direct CCF, indirect CCF tends to occur in an older age group (mean from 50 to 69 years of age), and women constitute 60–90% of all indirect CCF cases. Vascular changes in a variety of systemic conditions such as postmenopausal hormonal changes, pregnancy, hypertension, and atherosclerosis are hypothesized to predispose patients to the development of indirect CCFs.

Clinical manifestations of indirect CCFs include red eye, discomfort, ocular hypertension, or diplopia. Eye findings include engorged, "corkscrew" episcleral vessels from arterialized venous blood, ocular hypertension, and ophthalmoparesis. Less common presentations of indirect CCFs include headache, pulse-synchronous tinnitus, vision loss, and venous stasis retinopathy [28, 30, 31]. When indirect CCFs drain posteriorly to the superior or inferior petrosal sinuses, they may be asymptomatic or manifest as isolated cranial nerve palsies; symptoms and signs of orbital congestion become noticeable when indirect CCFs change its drainage from posterior to anterior draining indirect CCFs [32–34].

Importantly, significantly elevated venous pressure in the cavernous sinus may be transmitted retrograde to the cortical veins (cortical venous drainage), resulting in hemorrhagic venous infarction [35]. Cortical venous drainage may lead to severe neurological dysfunction such as hemimotor or hemisensory deficits, necessitating prompt intervention to close the arterial venous shunt. Among various clinical manifestations, the presence of bilateral orbital signs and a postauricular bruit was found to have the most predictive value of cortical venous drainage [36].

Radiographic Features

The most prominent radiographic feature of direct CCF and indirect CCFs on computed tomography (CT) or magnetic resonance imaging (MRI) is a dilated superior ophthalmic vein, although enlargement of the EOMs, abnormal cavernous sinus flow voids, and sometimes engorgement of the cavernous sinus with a convexity of the lateral wall can also be observed. These are best observed on thin section MRI [37].

CT angiogram and MR angiogram can add additional information [38, 39]. Orbital ultrasonography may also provide sensitive and reliable measurement by demonstrating dilatation and arterialization of flow of the superior ophthalmic vein but cannot give any information on posterior cortical venous drainage [40, 41].

A high index of suspicion should be maintained in patients with symptomatology as above, and a prompt imaging study of the brain and orbit using CT and/or magnetic resonance imaging should be done for screening. Catheter angiography remains the only definitive study to confirm or eliminate the diagnosis. Dural AVFs can be very difficult to diagnose with noninvasive imaging and are sometimes recognized only on catheter angiography.

Management of CCF

Spontaneous resolution of indirect CCFs due to venous thrombosis either unprovoked or after angiography has been reported in 20–60% of indirect CCFs, some in the literature [42, 43]. Spontaneous resolution of direct CCF has also been reported but is rarer [44, 45]. A nonsurgical management of indirect CCFs is carotid-jugular compression. The compression entails intermittent, seconds to a few minutes of compression of the ipsilateral cervical carotid artery and internal jugular vein using the contralateral hand for a period of a few weeks to a couple of months [42, 46, 47]. This maneuver should be considered in patients whose symptoms are too mild to warrant immediate surgical intervention or in those whose age or systemic comorbidities predispose them to higher surgical complications. Close follow-up may be indicated to evaluate for the development of cortical venous drainage (which is correlated with higher risk of development of hemorrhagic complications or venous infarcts) [35]. Indications for treatment include persistently elevated intraocular pressure, visual deterioration due to retina or optic nerve ischemia, severe proptosis, symptomatic ocular deviation with diplopia, exposure keratopathy, severe pain, and/or intolerable bruit.

A conservative approach is not likely to succeed in cases of direct carotid-cavernous fistulas. Resolution without recurrence has been described in only 17% of attempted cases [47].

Endovascular Treatment

Endovascular intervention has replaced intracranial surgery and is the treatment of choice when intervention is indicated. Endovascular treatment is typically the first-line approach for direct carotid-cavernous fistulas [48–50]. Compared to carotid artery surgery (trapping or ligation), endovascular intervention has significant lower risk of complications especially of cerebral ischemia. The objective of the endovascular treatment is to eliminate the arteriovenous carotid-cavernous shunting. This leads to normalization of the venous pressures, reversing the ophthalmic and leptomeningeal venous retrograde flow and engorgement, as well as symptoms related to vascular steal.

The evaluation for feeding pedicles during catheter-based angiography is made through bilateral common, internal, and external carotid artery iodinated contrast injections. The endovascular therapy aiming to obliterate the fistulous connections may be performed through either trans-arterial or transvenous routes, and it is based on the arterial and venous angioarchitecture and flow patterns. In direct fistulas, trans-arterial approach via the ipsilateral femoral artery then up through the internal carotid artery into the fistula ending into the cavernous sinus is often used. Different embolic materials are then injected into the cavernous sinus through the microcatheter; these may include detachable coils, *n*-butyl cyanoacrylate (acrylic glue), or ethylene vinyl alcohol copolymer [51]. The material of choice may be influenced by the size of the fistula. In case of a large tear, flow diverting stent assistance may be used [52, 53]. The stents require the use of antiplatelet therapy postoperatively.

Transvenous embolization of direct fistulas may be performed; however, it is not preferred due to the risks of migration of embolic material

into the ICA. For direct CCFs, overall endovascular occlusion rates have been reported to be between 55% and 99% with low <1% mortality [54–57], and the morbidity was described to be as high as 10–40%. In indirect CCFs, trans-arterial embolization is technically difficult due to the small size and multiplicity of feeders. Therefore, transvenous embolization of CCF is the preferred approach for indirect CCFs. Recent literature on endovascular approach promises high success rates of over 90%, with low complication rates ranging between 2.3% and 5% [50, 58]. The cavernous sinus is most commonly accessed through the inferior petrosal sinus; however, there are multiple other endovenous approaches including catheterization of the superior petrosal sinus or the facial vein takeoff from the internal jugular vein or the arterialized dilated superior ophthalmic vein can be directly accessed via an eyelid crease incision [59, 60].

Regardless of the access route, the microcatheter is optimally positioned within the cavernous sinus (typically more anteriorly, close to the proximal aspect of the superior ophthalmic vein) and embolization can ensue. Commonly used options are coiling and/or embolization with ethylene vinyl alcohol copolymer (Onyx). The sinus is filled with coils and/or liquid embolics until there is complete distribution of the embolic material within the cavernous cavity, and no early venous drainage is observed through arterial runs. Ocular symptoms tend to improve over the following hours. Paradoxical worsening of the symptoms has been described but tends to be transient [61]. Coil overpacking or direct liquid embolic local effects on nerves may generate posttreatment cranial nerve deficits (which commonly improve) [62].

Successful angiographic closure was achieved in 93% of direct and 92% of indirect fistulas. Multiple treatments were required in 33% of direct and 16% of indirect fistulas [63]. When an endovascular approach is not feasible or has been unsuccessful, stereotactic radiosurgery (SRS) may be considered for treatment of a dural CCF. Radiosurgery has been demonstrated in case series to be effective with 90% success at 1–2 years [64, 65]; however, the latency period for the obliteration of the fistula and symptom improvement is typically of several months; hence, it is not appropriate if there is risk of acute visual or neurologic worsening. It may have an important role for incompletely treated indirect fistulas. Using a therapeutic radiation dose of about 20 Gy, SRS induces an injury of the targeted vessel, thus obliterating the vessel lumen.

Illustrative Case 1

A 57-year-old woman presented with diplopia and evidence of lateral rectus paresis. Subsequently she developed chemosis, prominent episcleral vessels, and increased intraocular pressures involving the left eye (Fig. 15.3a). A MRI orbit with contrast showed prominent cavernous sinus and enlarged superior ophthalmic vein in left orbit (Fig. 15.3b) which showed arterialization of flow on MRA (Fig. 15.3c). Conventional angiography confirmed a Barrow Type C indirect fistula which means it only had filling from a deep branch of the internal maxillary artery ultimately fed by the external carotid artery. There was no filling from the internal carotid. Early arterial phase filling of the cavernous sinus and the arterialized superior ophthalmic veins were clearly demonstrated (Fig. 15.3d, e). The microcatheter was advanced through the femoral vein into the left jugular vein, then into the enlarged left facial vein, and ultimately through the superior ophthalmic vein into the cavernous sinus. This approach was used to coil embolize the fistula. She did well post intervention and showed resolution of episcleral congestion and normalization of her extraocular motility (Fig. 15.3f).

Idiopathic Intracranial Hypertension

Introduction

Idiopathic intracranial hypertension (IIH), also called pseudotumor cerebri or benign intracranial hypertension, refers to a condition of elevated

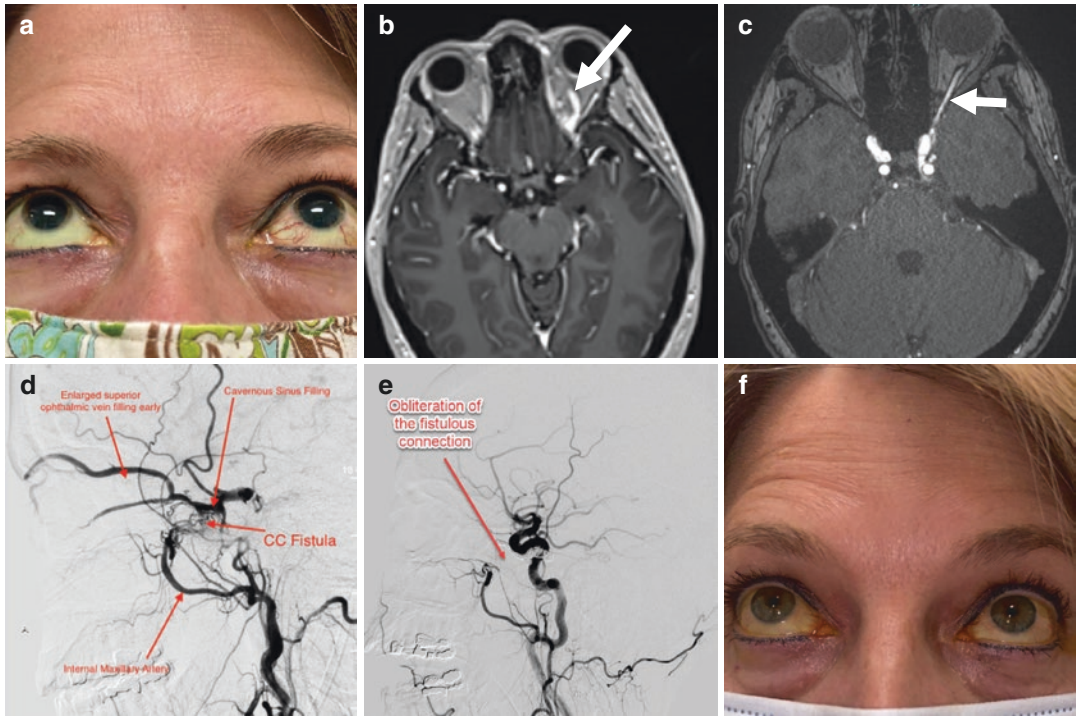

Fig. 15.3 (**a**) Clinical photo demonstrating prominent episcleral vessels of the left eye compared to normal appearance of right conjunctiva and sclera. (**b**) MRI orbit with contrast demonstrating enlarged superior ophthalmic vein in left orbit *(white arrowhead).* (**c**) MRA of the head with contrast demonstrating arterialization of the superior ophthalmic vein (*white arrowhead*). (**d**) Lateral angiogram projections that show the caroticocavernous (CC) fistula with early cavernous sinus filling from a deep branch of the internal maxillary artery and early filling of enlarged superior ophthalmic vein. (**e**) Post coil embolization angiogram showing obliteration of the fistulous connection as shown by lack of early arterial phase filling of the cavernous sinus. (**f**) Clinical photo after coil embolization that shows resolution of episcleral congestion left eye. (Panel **d** and **e** angiogram images are courtesy of Dr. Rano Chatterjee, Washington University School of Medicine, Department of Neuroradiology)

intracranial pressure unrelated to a space-occupying lesion, cerebral venous thrombosis, meningitis, or hydrocephalus. IIH has a predilection for obese women of child-bearing age, although it can occur in children, at older age, and in males [66–69]. IIH has been associated with a variety of medications including antibiotics (tetracycline, minocycline, doxycycline, and nalidixic acid), growth hormone, lithium, retinoids (both topical and oral), Lupron [70], and cyclosporine. Obstructive sleep apnea and recent weight gain may also contribute to an elevated intracranial pressure [71].

A number of mechanisms are thought to contribute to the development of IIH, including increased cerebrospinal fluid production, reduced cerebrospinal fluid absorption, the influence of hormones, abnormal vitamin A metabolism, as well as elevated cerebral venous pressure. Still to be explored is the role of the newly named "glymphatic system" in the pathogenesis of IIH [72, 73]. The role of elevated intracranial dural venous pressure in the pathophysiology of IIH has gained increasing attention, as a potentially treatable cause of IIH. Although stenosis of the transverse and sigmoid sinus is a common radiographic finding in IIH [74], it is unclear whether dural sinus stenosis is a cause of elevated intracranial pressure or a consequence of chronic compression of dural venous sinuses from persistently elevated intracranial pressure. Regardless of the etiology, increased resistance in cerebral venous

outflow seems to be the common final pathway in the pathophysiology of IIH, suggested by elevated manometry measurements of the prestenotic vs. poststenotic pressure gradient [75–86].

Clinical Manifestation

The typical presentation of IIH includes headache, pulse-synchronous tinnitus, with varying degrees of vision loss, and papilledema. Headache occurs in about 90% of IIH patients [87]. Prior studies suggested headaches associated with IIH most often presented as pain in a nerve root distribution or as retro-ocular pain with eye movement [88, 89]. More recently, the understanding has changed, and migraine is recognized as the predominant phenotype [90]. Pulse-synchronous tinnitus, described as a "whooshing" sound synchronized with the heartbeat, is more specific for IIH if present. Patients may complain of transient visual obscurations and episodic and severe vision loss in both eyes lasting for seconds with complete recovery usually associated with activities that increase central venous pressure (such as Valsalva maneuver) or decrease systemic perfusion pressure (transition from supine or sitting to the upright position). Papilledema, manifested as hyperemia and elevation of the optic disc, engorgement of the central retinal veins, and obscuration of the central retinal vessels on the disc often with disc hemorrhage and exudates, is usually bilateral and symmetrical (Fig. 15.4). Most patients with IIH have mild vision loss that is reversible after appropriate treatment, although permanent vision loss can occur in about 25% of patients [89]. A small proportion of patients (2–3%) with IIH present with fulminant visual loss over days [91], necessitating aggressive intervention to salvage vision.

Investigation and Diagnosis

The diagnostic criteria of IIH were created by Dandy in 1937 and were formulated by Friedman in 2002 [92]. In 2013, Friedman et al. proposed a revision to the diagnostic criteria that separated patients into two groups: those with papilledema and those without papilledema [93]. In both groups, the revised diagnostic criteria emphasize a normal neurological exam with the exception of cranial nerve abnormalities, normal cerebrospinal fluid composition, as well as brain imaging study using MRI and MR venography (MRV) to rule out intracranial pathologies that may cause secondary intracranial hypertension. The diagnosis is confirmed in patients who also demonstrate cerebrospinal opening pressure greater than 250 mm H2O and normal CSF analysis and papilledema. In the absence of papilledema, the diagnosis may be confirmed by the presence of a unilateral or bilateral abducens nerve palsy or at least three of the characteristic neuroimaging findings associated with IIH [93]. Diagnostic procedures include MRI and MRV of the brain and a lumbar puncture. Lumbar puncture provides information about the opening pressure as well as cerebrospinal fluid profile; the latter is essential in excluding secondary causes such as inflammation or infection. When performed without anesthesia with patients lying in the lateral decubitus position, the intracranial pressure in normal adults ranges from 100 to 250 mmH2O [92, 94]. MRI and MRV of the brain provide information to rule out intracranial pathologies such as space-occupying lesions, hydrocephalus, Chiari malformation, and cerebral venous thrombosis. The radiographic findings suggesting IIH include posterior globe flattening, optic nerve sheath distension, empty sella, and transverse venous sinus stenosis [95]. One study using a specially designed MRV protocol found stenosis in the transverse and sigmoid sinuses to be both sensitive and specific for IIH [74].

Management

The severity of the vision loss in patients with IIH is the main determinant for treatment strategy. Treatment of patients with severe headache but intact visual function and mild papilledema is more variable across practitioners. Lumbar punc-

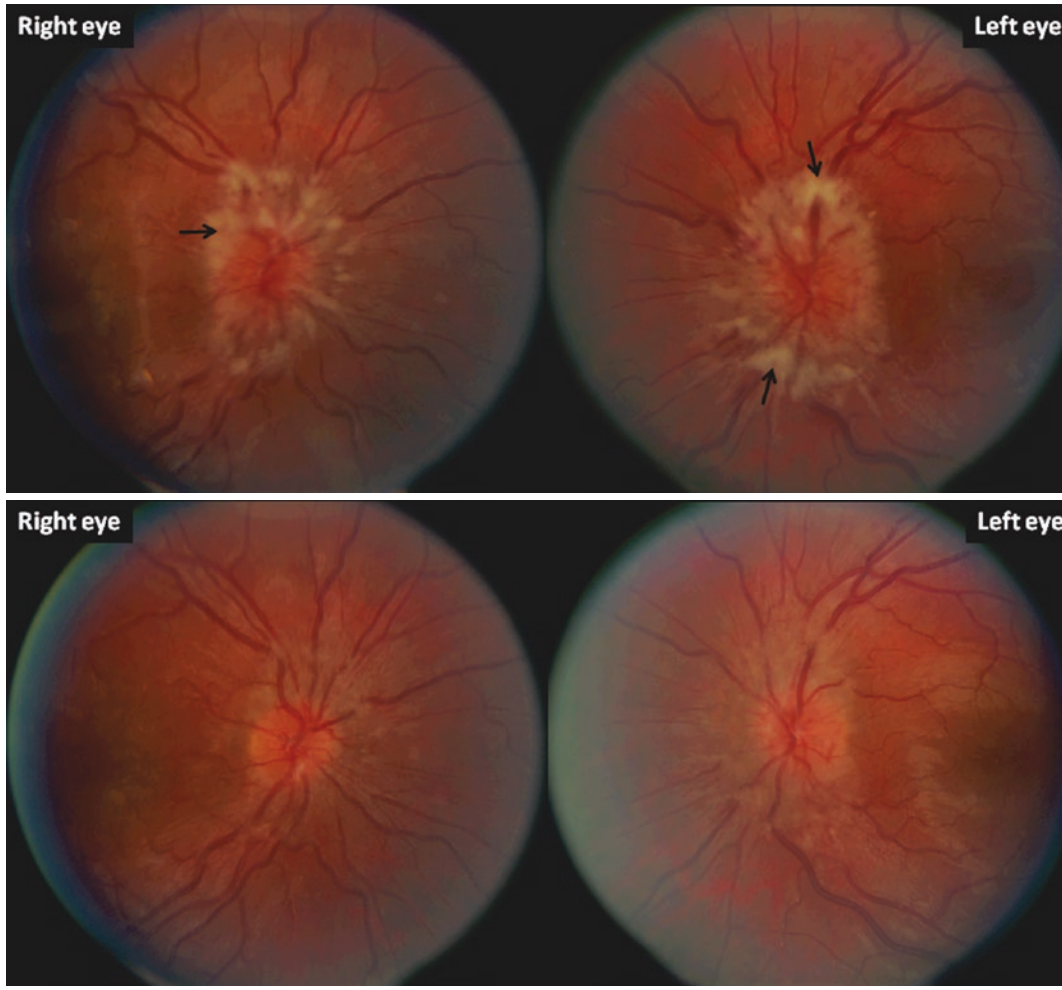

Fig. 15.4 Bilateral papilledema in a 26-year-old lady with concurrent right sixth nerve palsy. Note the elevation of the optic disc, engorgement of the central retinal veins, and obscuration of the retina vessels both on the disc and as the vessels exit the disc margin (*top*). There are disc hemorrhages in the left eye. There are nerve fiber layer infarcts reflected by the white areas (*arrows*). The patient's visual acuity was 20/40 in the right eye and 20/60 in the left eye. The patient had been on oral minocycline for 2 months for acne treatment prior to presentation. Minocycline was discontinued, and she was given acetazolamide 500 mg twice daily. One month later, both six nerve palsy and papilledema improved (*bottom*)

ture serves as one of the mainstays of diagnosis [96]. While sometimes resulting in relief of headaches, repeated lumbar punctures are not recommended as a treatment methodology.

Weight loss serves as an important step in the management of IIH, and successful reduction of body weight of 5–10% may have a significant impact on the evolution of both headache and papilledema [97, 98]. Pharmacologic treatment commonly includes carbonic anhydrase inhibitor and topiramate, or methazolamide or furosemide can be used when acetazolamide is poorly tolerated [99]. Prospective data from the Idiopathic Intracranial Hypertension Treatment Trial (IIHT) showed that treatment with acetazolamide in conjunction with weight loss resulted in statistically significant improvement in visual field function in patients with IIH and mild visual loss

compared to placebo and weight loss [100]. One must be aware that patients diagnosed with IIH may suffer from a headache disorder above and beyond that related to the elevated intracranial pressure, given the high predilection of headache disorder in the same demographic population.

When there is imminent visual loss, surgical intervention may be required. The current surgical options for treatment of IIH are optic nerve sheath fenestration, CSF diversion (lumboperitoneal or ventriculoperitoneal shunt), or venous sinus stenting. Currently, there are no evidence-based guidelines regarding choosing a surgical procedure to treat severe IIH, so the therapy used is based on local availability and expertise. Optic nerve sheath fenestration creates a window or multiple slits on the intraorbital segment of the optic nerve sheath behind the globe to release cerebrospinal pressure [101]. Optic nerve sheath fenestration is generally regarded as a low-risk procedure, although serious complications may rarely occur such as central retinal artery occlusion, resulting in profound loss of vision. Lumboperitoneal and ventriculoperitoneal shunts divert cerebrospinal fluid from the spinal canal or cerebral ventricle into the abdomen via a catheter to lower the intracranial pressure. Image-guided shunt placement may result in improved shunt placement accuracy but does not affect long-term shunt survival [102, 103]. This allows treatment of headache and leads to stabilization or improvement of visual acuity and visual fields. Shunting may also decrease average retinal nerve fiber layer thickness and improve Frisen papilledema grade [104]. However, revision surgery is frequently necessary, and other complications (as low-pressure headache, infection, arachnoiditis of nerve roots) might develop [105, 106]. Complication rates and shunt removal rates do not differ between lumboperitoneal and ventriculoperitoneal shunts [107, 108]. Venous sinus stenting is another emerging treatment modality that addresses the transverse-sigmoid sinus stenosis and resulting transvenous pressure gradient commonly seen in patients with IIH. This is discussed below.

Endovascular Treatment

Venous sinus stenting is a more recent endovascular treatment option for IIH that has been used since the early 2000s [108].

Venous sinus stenosis appears to result from increased intracranial pressure, thus decreasing CSF resorption into the venous system and causing worsening intracranial pressure. However, there is still some discussion as to whether venous sinus stenosis is the cause or result of elevated intracranial pressure, and the exact mechanism has not yet been definitively elucidated [109]. Patients with IIH were found to have substantial bilateral sinovenous stenosis in 27 of 29 (93%) patients with IIH versus 4 of 59 (6%) in control patients by MR gadolinium-enhanced venography [74].

Venous sinus stenting may be considered in patients who demonstrate a pressure gradient across the respective stenotic sinus. The overall rate of an elevated cerebral venous pressure gradient in patients with IIH is 35% which is determined on catheter venography after lumbar puncture [110]. Current evidence suggests a promising role of venous sinus stenting with significant improvements in papilledema, headache, and pulsatile tinnitus and improved or stabilized vision with few 1-year or even late failures [77, 106, 111]. However, though complications are relatively rare, they can be severe [80, 111, 112].

A meta-analysis from 2019 included 474 patients with IIH that were treated with venous sinus stenting. The overall rate of improvement of headaches was reported to be 79.6%. Papilledema was reported to be improved in 93.7%, and pulsatile tinnitus improved in 90.3% of patients. The rate of major complications was 1.9% [111].

A systematic review published in 2020 included 47 studies that represented 825 patients in total with follow-up ranging from 0 to 136 months. Resolution or improvement was observed in 87.1% of the cases with papilledema and 72.1% of the cases with headaches. Major complications occurred in 19 patients (2.3%) and

included subdural hematoma, intracerebral hematoma, subarachnoid hemorrhage, and obstructive hydrocephalus. Symptom relapse occurred in 25 patients (3.4%) and required restenting or supplemental intervention [111].

A review from 2018 of 32 studies that included 186 patients found that higher mean pressure gradients (22.8 ± 11.5 mm Hg vs. 17.4 ± 8.0 mm Hg, $p = 0.033$) and higher changes in pressure gradients (19.4 ± 10.0 mm Hg vs. 12.0 ± 6.0 mm Hg, $p = 0.006$) after stent placement were associated with improved clinical outcomes. After controlling for age, sex, body mass index, CSF opening pressure, pre- and post-stent pressure gradient, the change in pressure gradient was found to be an independent predictor of favorable outcomes ($p = 0.028$) [109].

Periprocedural Steps

The first step in endovascular treatment involves angiographic confirmation of dural sinus stenosis and measurement of the trans-stenotic pressure gradient (typically ≥8 or 10 mmHg is considered clinically significant [77, 113]. Guide catheter navigation and venous sinus stenting can be painful, and general anesthesia is generally used. However, several recent studies have demonstrated that general anesthesia results in significant differences in venous sinus pressure measurements [114, 115]. Based on these findings, conscious sedation may be preferred to general anesthesia when pressure measurement is being conducted, with conversion to general anesthesia if the determination to stent is made [115]. Patients must be pretreated with aspirin and clopidogrel (3–5 days of 75 mg nightly or a 300 mg loading dose 24 h prior to stenting). Intraprocedurally, therapeutic heparin anticoagulation (ACT > 250 s) is recommended. Via an ipsilateral femoral or IJ vein, a 6F is inserted, and a properly sized self-expanding stent (or balloon-expandable) is navigated into the venous sinus stenosis. Pre- and post-angioplasty may be performed, but typically not necessary. Dual antiplatelet therapy should be maintained for at least 3 months.

In summary, stenting in appropriately selected patients with refractory IHH is showing a promising role.

Illustrative Case 2

A 37-year-old woman with a history of rheumatoid arthritis presented with worsening frequency/intensity of headaches and blurry vision. Exam revealed papilledema. MRI brain was normal, and MRV was suspicious of venous stenosis bilaterally. Lumbar puncture revealed opening pressure of 24 cm H_2O. Headache was refractory to acetazolamide and repeated therapeutic lumbar punctures. Her vision worsened, and she was referred for optic nerve fenestration. Despite this therapy, headaches persisted.

Cerebral venous manometry and possible angioplasty/stenting were requested. Stenosis of the right transverse-sigmoid junction was observed (Fig. 15.5a, b). The mean venous pressure in the superior sagittal sinus was 41 mmHg (Fig. 15.5c), torcula 39 mmHg, transverse sinus (distal to stenosis) 36 mmHg, sigmoid sinus (proximal to stenosis) 29 mmHg, and jugular bulb 18 mmHg. Due to the gradient of 23 mmHg, the decision was made to intervene endovascularly. Angioplasty with a 6 × 40 mm Savvy balloon was performed. This was followed by the deployment of an 8 × 20 mm precise self-expandable stent. Due to the presence of stenosis immediately distal to the stent, an overlapping 8 × 30 mm Protégé self-expandable stent was placed. A good angiographic result was noted, and pressures in the superior sagittal sinus decreased to 22 mmHg, transverse sinus to 21 mmHg, and jugular bulb to 19 mmHg. Chronic daily headache and visual blurriness resolved within weeks. Follow-up angiogram at 2 months revealed patency of the stents. At 1 year, the optic disks were normal.

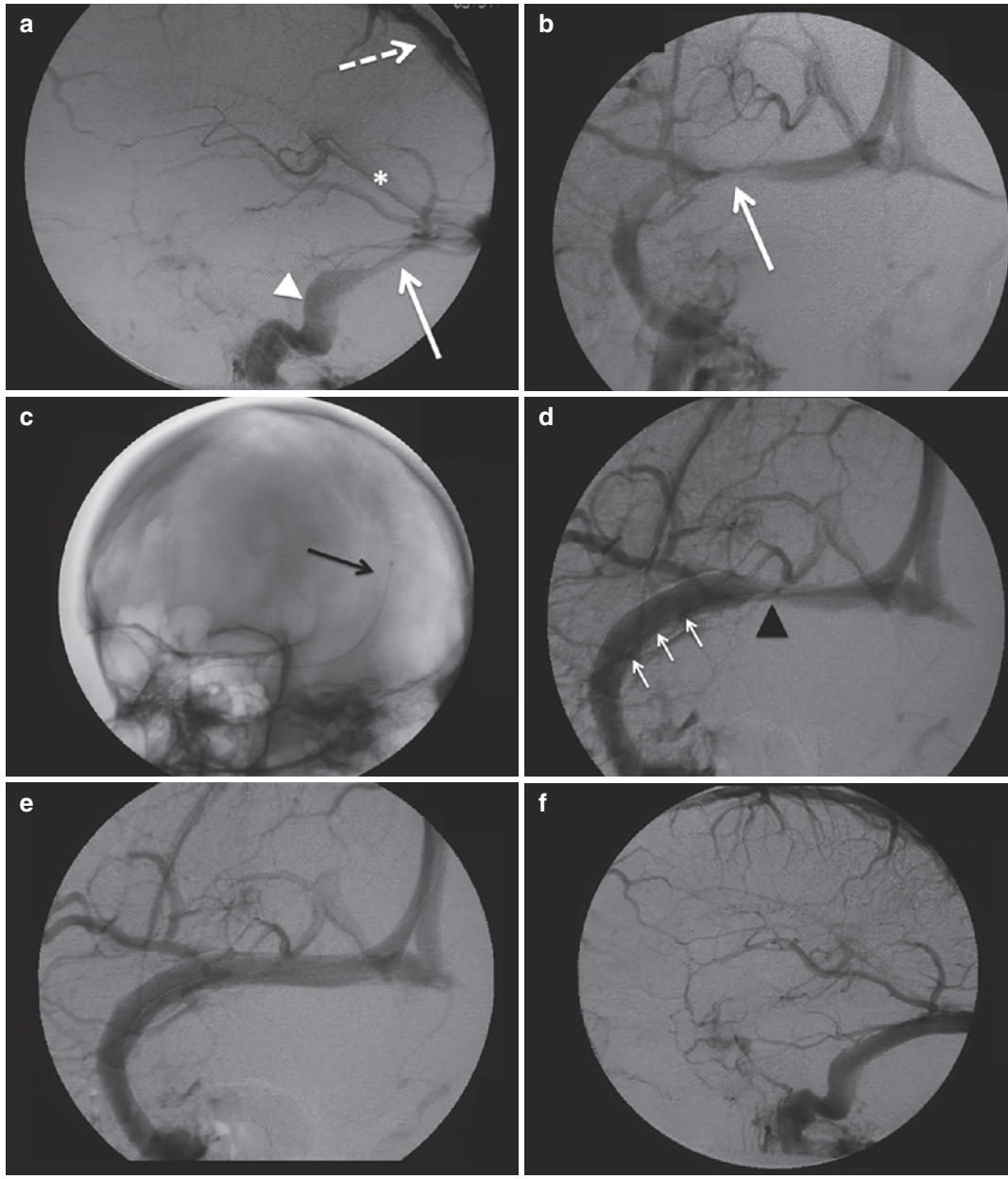

Fig. 15.5 (**a**) Lateral venous phase of arterial angiogram revealing stenosis of the transverse-sigmoid junction (*white arrow*). The sigmoid sinus is normal (*white arrowhead*), and the superior sagittal (*dashed arrow*) and straight sinuses are observed (*asterisk*). (**b**) An anterior–posterior projection reveals stenosis of the transverse-sigmoid junction (*white arrow*). (**c**) Oblique projection revealing the microcatheter positioned in the superior sagittal sinus. (**d**) A stent is depicted (*small white arrows*), and a residual segment of stenosis noted (*black arrowhead*). (**e**, **f**) Fully patent sinuses

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