Letters to the Editor

Papilledema: A Comprehensive Assessment

Sir,

Papilledema is characterized by optic disc edema when there is an increased intracranial pressure (ICP). It must be distinguished from pseudopapilledema (optic disc drusen, hyperopia) and other causes of optic disc swelling (intraocular inflammation, central retinal vein occlusion, optic neuritis, hypertensive disc edema, ischemic optic neuropathy, infiltrative tumors, compressive optic neuropathy, and scleritis). As it consists of an alarming sign for intracranial entities, efforts must be taken to make a promptly diagnosis and management.

Unilateral optic disc edema is commonly observed in inflammation, ischemia, compression, or infiltration of the optic nerve. However, up to 2% of cases of papilledema may be unilateral or very asymmetric.[1] In these cases, ocular and orbital conditions must be investigated.[2] Recently, a case of unilateral papilledema associated with cerebral venous sinus thrombosis was described, evidencing a rare association in this challenging entity.[3] Optic nerve sheath anomalies (preventing the transmission of the raised cerebral spinal fluid [CSF]) and lamina cribrosa changes (increased collagen and decreased elasticity with aging) were hypothesized to explain unilateral papilledemas.[4]

CSF opening pressure (which typically ranges from 10 to 25 cm of water) is the gold standard tool to diagnose increased ICP. Expansive lesions (tumor, hemorrhage), decreased absorption of CSF (hydrocephalus, venous outflow obstruction), increased CSF production (choroid plexus tumor) or a pseudotumor cerebri (idiopathic) can result in high CSF-increased ICP.[5] Symptoms include headaches (worse in the morning), transient obscuration of vision (when raising to stand), and nausea and vomiting. It can also lead to cranial nerve palsies (commonly abducens palsy).[6]

Suspicious patients should be asked for pregnancy, systemic diseases, and use of medications. Light and accommodation pupil responses must be tested. Fundoscopy ranges from C-shaped halo with a temporal gap in early phases. Circumferential retinochoroidal folds surround the disc, called Paton’s lines, are usually temporal and may be present. Late phases show elevation of the nerve head, obscuration of a segment of a major blood vessel, and obliteration of the optic cup. The absence of spontaneous venous pulsations also suggests increased ICP.

When there is a hypothesis of papilledema, the first approach is imaging with MRI, to rule out expansive lesions. Flattening of the sclera near the optic nerve and a raised area at the disc head may be observed. Whenever there is no contraindication, lumbar puncture with opening pressure must be performed to confirm increased ICP, and CSF must be sent for cytology and culture.[7]

Despite ICP is treated primarily to prevent herniation and to preserve cerebral blood flow, persistent papilledema may lead to optic nerve atrophy and permanent vision loss. Interruption of causative agents should be done promptly. Diuretics are the first line of treatment, can reduce ICP by increasing CSF absorption or decreasing its production, and usually are well tolerated. Surgical procedures include creation of ventriculoperitoneal or lumboperitoneal shunts, which normalizes ICP by increasing CSF drainage, and should be considered when medical therapy fails. In selected cases, when vision is threatened, optic nerve sheath fenestration may be performed to reach its proper decompression.[8]

Currently, there is a lack of new medications to treat specifically increased ICP. Improvement in neuroimaging modalities could make a diagnosis of papilledema progressive safer, faster, and easier. However, direct fundoscopy remains as an important tool for the diagnosis and to guide the treatment in those cases.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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REFERENCES

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