Case Report

Unilateral Papilledema with Bilateral Optic Nerve Sheath Distension: A Case Report

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Abstract

Bilateral edematous optic disc swelling from papilledema is caused by elevated intracranial pressure (ICP). Idiopathic intracranial hypertension (IIH), a clinical syndrome with elevated ICP of unclear etiology, is a frequent cause of this condition. IIH typically affects obese middle-aged females. Papilledema usually has a fairly symmetrical bilateral pattern. Unilateral papilledema is a rare disorder that must be detected early to avoid optic nerve damage. However, the etiology of unilateral papilledema remains unclear. Based on bilateral optic nerve sheath diameter measurements, we aimed to find an explanation for the unilaterality in this rare case.

Keywords: Idiopathic intracranial hypertension, optic nerve sheath diameter, unilateral papilledema

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Introduction

Papilledema is bilateral and nearly symmetrical optic disc swelling attributed to increased intracranial pressure (ICP).[1] One of the common causes of ICP is idiopathic intracranial hypertension (IIH), a clinical syndrome of increased ICP of unknown etiology that usually occurs in obese middle-aged females.[2] Unilateral papilledema is a rare condition, reported in 4% of all IIH patients, and may be misdiagnosed as local ocular pathology, making IIH diagnosis difficult.[3,4,5] In the presented case, we attempted to assess optic nerve swelling using fundus photography, optical coherence tomography (OCT), and ultrasound (US) measurement of optic nerve sheath diameter (ONSD) to establish the diagnosis despite the unilateral condition. Lumbar puncture with measurement of cerebrospinal fluid (CSF) opening pressure was done to confirm increased ICP and confirm the diagnosis of unilateral papilledema. The cause of unilateral papilledema is still unclear. Based on the data obtained, including bilateral increased ONSD, we aimed to identify the etiology of the unilaterality in these rare cases.

Case Reports

A 35-year-old obese woman presented to our clinic with transient visual obscuration in both eyes that persisted for a few seconds. The patient denied any other visual complaints, redness, itching, ocular pain, or discharge. She complained of frequent one-sided headaches but denied having diplopia, tinnitus, neck stiffness, and weakness or numbness of the limbs. The patient reported no allergy or drug reactions, and she was a non-smoker, non-alcoholic, non-diabetic, and non-hypertensive, with no history of using oral contraceptives, corticosteroids, or other drugs or positive family history of autoimmune or neurological diseases.

The patient had no complaints of chest pain or shortness of breath. On physical examination, she was found to be obese (body mass index of 30 kg/m^2) with no other detected abnormalities. Body temperature and blood pressure (110/70 mmHg) were normal. Laboratory results indicated normal cell counts and random blood glucose, with no vitamin or iron deficiency.

Visual and optic nerve function tests revealed bilateral 20/20 visual acuity and normal color vision and contrast sensitivity with no evidence of red color desaturation. Pupillary reaction was normal on both sides. Anterior segment examination was normal, with no evidence of conjunctival injection, congestion, cells, or flares in the anterior chamber. Ocular motility was normal and painless in all directions. Intraocular pressure (IOP) was within normal range (18 mmHg). There were no signs of proptosis or eyelid swelling.

Fundus examination revealed a normal right fundus with no disc swelling, while the left eye exhibited disc swelling at both the nasal and temporal margins with circumferential halos and obscuration of some blood vessels upon leaving the disc. Paton lines were observed with absent venous pulsations bilaterally. There was no evidence of vasculitis, retinitis, vitritis, or optociliary shunts. Fundus photography and fluorescein angiography of the right eye showed well-defined disc margins and no evidence of optic disc swelling, early dilated capillaries, or late dye leakage. The left eye had a swollen disc with blurred margins, partially obliterated cupping, Paton lines, and no evidence of anomalous blood vessel bifurcations. Fundus fluorescein angiography showed early dilated capillaries with late optic disc leakage (hot disc; Figure 1).

OCT showed a completely normal right disc with preserved cupping, no evidence of peripapillary fluid, and normal retinal nerve fiber thickness (RNFLT; average 109 µm) with normal double hump pattern and minimum rim width (MRW). However, OCT of the left eye showed peripapillary fluid with hyporeflective triangles, a partially obliterated disc cup, increased RNFLT (average 179 µm), an elevated double hump curve, and a thickened MRW with an elevated curve. Macular scanning revealed no evidence of macular thickening or macular intraretinal fluid. Bilateral enhanced depth image OCT (EDI-OCT) demonstrated no evidence of disc drusen (Figure 2).

Blue-light fundus autofluorescence revealed no optic disc drusen, well-defined disc margins in the right eye, and blurred margins in the left eye (Figure 3). Furthermore, US measurements of ONSD demonstrated thickening bilaterally, with a right ONSD of 6.61 mm and a left ONSD of 6.9 mm (Figure 4). Neuroimaging including magnetic resonance imaging (MRI) and magnetic resonance venography revealed no evidence of space-occupying lesions, inflammation, transverse sinus stenosis, or occlusion. However, MRI indicated flattening of the globe and tortuous optic nerve in the left eye with prominent CSF around the optic nerve bilaterally (Figure 5).

The patient was then eligible to undergo lumbar puncture and was referred to the neurological department. The opening pressure was 35 cmH_2O and CSF analysis was normal. Accordingly, the case was diagnosed as IIH according to the Dandy criteria. The patient received 500 mg of acetazolamide twice daily and 25 mg topiramate once daily with diet therapy, and instructions for weight reduction and follow-up after one month were recommended.

After 1 month, the patient reported resolution of the visual obscuration and a decrease in headache attacks, which disappeared completely after 4 months. The results were confirmed by fundus photography, which showed the disappearance of previously observed peripapillary intraretinal fluid. OCT revealed a significant reduction in RNFLT in the left eye with a reduction in the double hump curve and MRW (Figure 6). However, there was only a slight decrease in ONSD (average 6.2 mm).

Discussion

Unilateral optic disc swelling may be caused by several conditions, including anterior ischemic optic neuropathy (AION), optic neuritis, disc drusen, compressive and infiltrative optic neuropathy, disc tumors, papillophlebitis, diabetic papillopathy, neuroretinitis, and rarely, papilledema.[6] Whenever confronted with a case of unilateral papilledema, it is recommended to obtain a comprehensive medical and familial history, conduct a thorough evaluation of visual function and pupillary reactions, and consult with an ophthalmologist to assess IOP, perform slit-lamp and fundus examinations, and utilize diagnostic techniques including OCT, fundus photos and autofluorescence

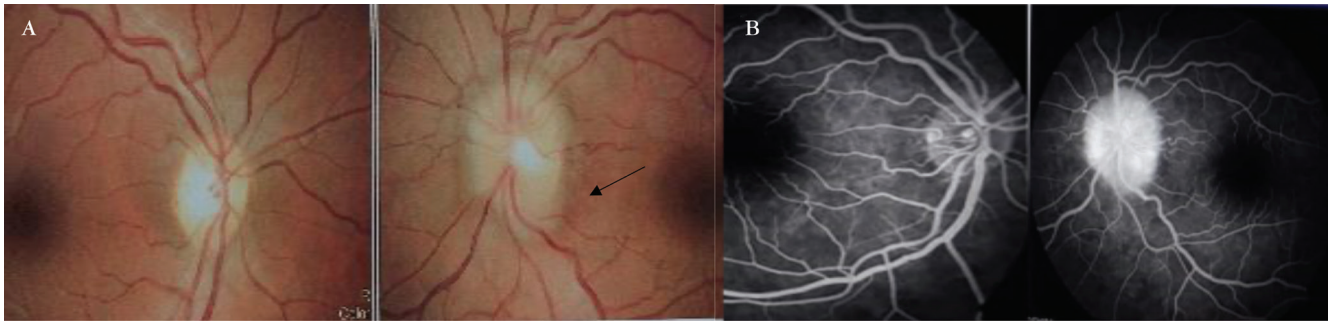

Figure 1. (A) Fundus photography shows the right normal optic disc and left swollen disc with Paton lines (black arrow). (B) Fluorescein angiography shows no evidence of late leakage on the right and significant late disc leakage on the left

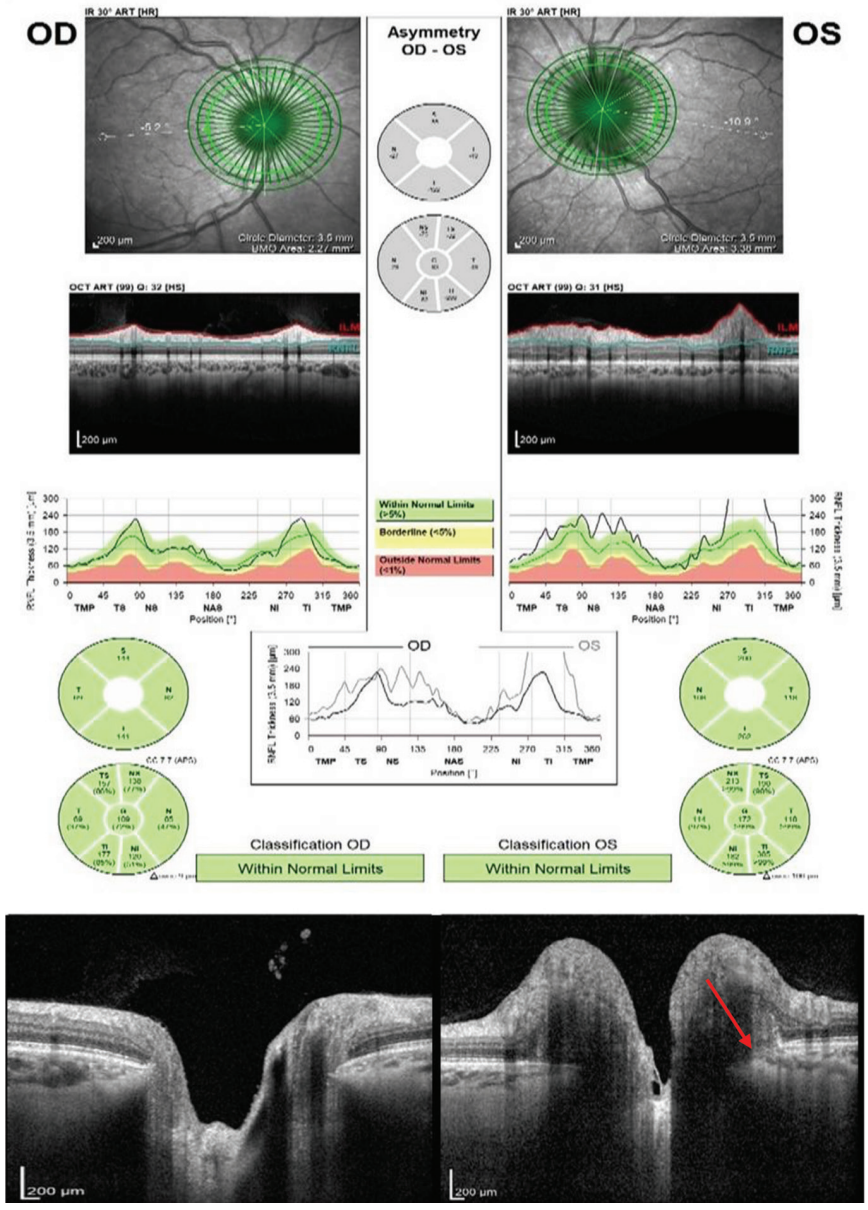

Figure 2. Optical coherence tomography showing retinal nerve fiber thickness in both eyes (top panels) and line scans (bottom panels) demonstrates a normal right eye and peripapillary fluid with a hyporeflective triangles and increased disc height in the left eye. Notice the superior displacement of Bruch's membrane described in papilledema (red arrow)

for excluding optic disc drusen. MRI of the brain and orbits and venography should be performed to reveal compressive lesions, optic nerve tumors, space-occupying lesions, and signs of increased ICP, if present.[7]

In our case, normal visual acuity, color vision, and pupillary reactions ruled out AION while optic disc drusen and optic neuritis were excluded through blue-light autofluorescence and EDI-OCT. The patient was non-diabetic and non-hypertensive, with MRI showing no compressive lesions. However, ONSD measured by US indicated bilateral thickening, raising high suspicion of increased ICP. This was confirmed by elevated lumbar puncture and a good response to medical treatment, with complete resolution. In our case, we used different modalities to detect the unilaterality of the condition. Some prior case reports of unilateral papilledema depended only on fundoscopy, which may miss subtle edema in the other less edematous eye. Others only used OCT to assess optic disc edema.[8,9] Swinkin et al.[9] assessed optic nerve sheath distension using fundus photography, OCT optic nerve scanning, and MRI. To the best of our knowledge, our report is the first to use US to measure ONSD in a case of unilateral papilledema, demonstrating that the sheath was relatively dilated in the right eye despite appearing normal in all other modalities.

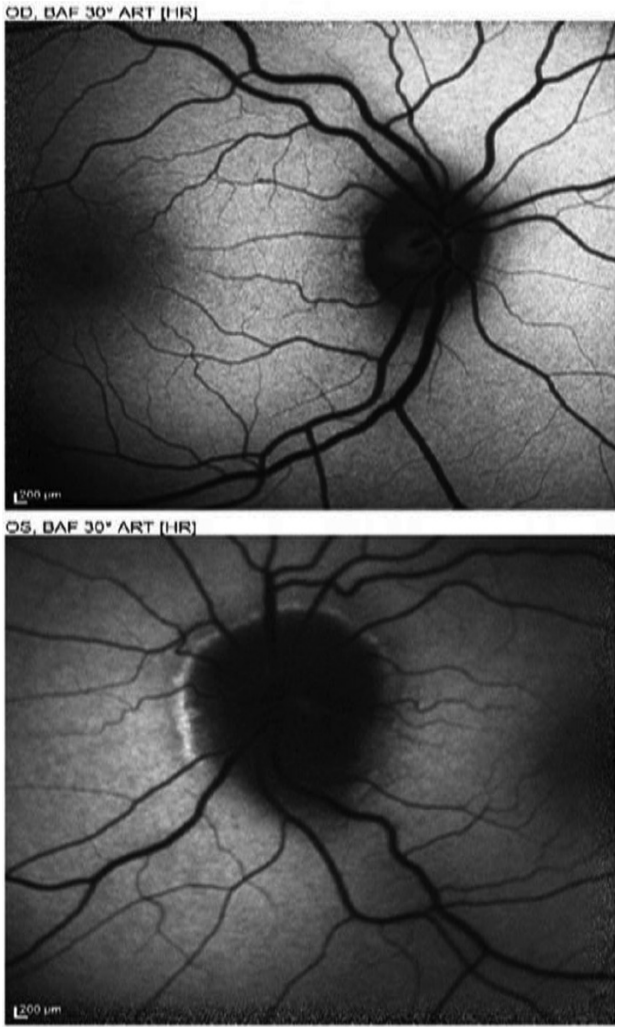

Figure 3. Blue autofluorescence shows well-defined disc margins in the right eye (top) and ill-defined margins in the left eye (bottom)

Measurement of ONSD using US is a generally accepted and documented procedure in the assessment of patients with increased ICP. Recently, it has been recommended, possibly routinely, in all cases suspected of IIH.[10] Nevertheless, its application is not very widespread, and in fact the modified Dandy criteria, which include clinical, laboratory, and radiographic data for the diagnosis of IIH, do not describe US or ONSD measurement. Regrettably, the B-scan has several drawbacks, including the blooming effect and distortions that can skew the results. To overcome that, we had to be cautious to average at the lowest feasible gain when utilizing mode B.[11,12]

There are hardly any situations in which unilateral papilledema is observed. In patients with pre-existing unilateral optic atrophy, only the normal eye experiences papilledema if ICP is later elevated, as in Foster-Kennedy syndrome. Differences in venous drainage between the eyes, differences in structural disc characteristics (including lamina cribrosa abnormalities), and unilateral highly myopic cases with significant differences in the anatomical path of the optic nerve are other proposed causes of unilateral papilledema documented in the literature.[4,13]

Many theories have been proposed to explain the occurrence of unilateral papilledema, but the reasons remain unclear. The relationship between CSF pressure, IOP, and systemic blood pressure is thought to play a role in papilledema occurrence. Whenever CSF pressure increases, IOP decreases, or perfusion pressure becomes lower. This is thought to cause axoplasmic flow stasis and optic disc edema.[14] Although the patient in our case showed bilateral normal IOP, she was non-hypertensive with normal systemic blood pressure. Accordingly, the only possible cause would be increased CSF pressure at the optic disc. However, ONSD was increased bilaterally and decreased after lumber puncture, indicating successive treatment and reduction in ICP.[15] Another theory was that a smaller bony canal on the side of the normal nerve might have a role in preventing transmission of CSF pressure along the optic nerve, resulting in less edema.[16] Conversely, Farrokhi et al.[17] found that the optic nerve bony

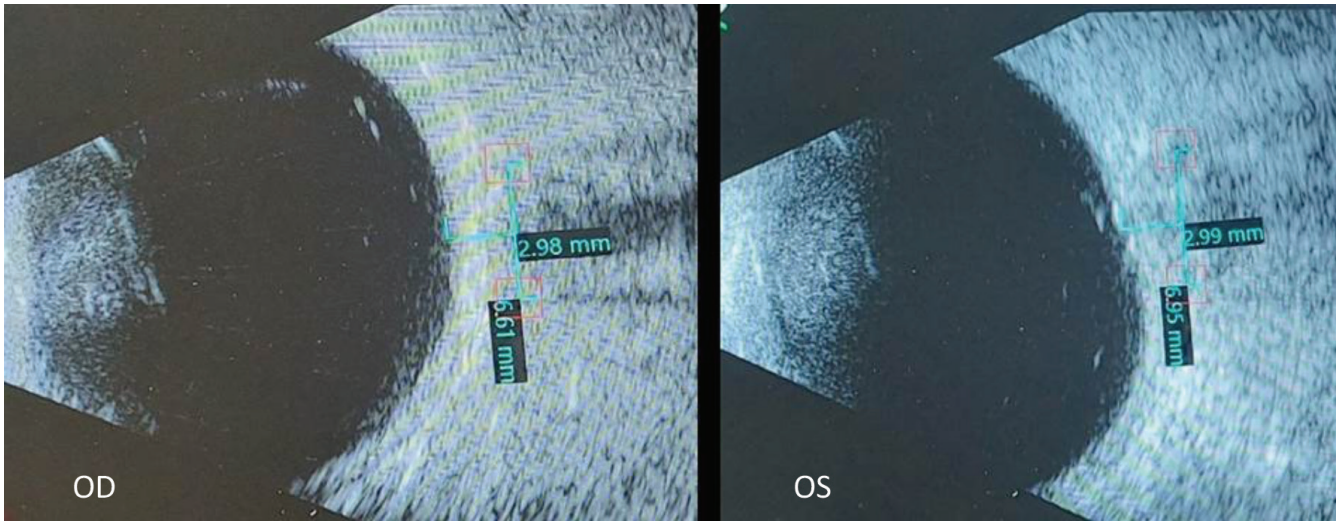

Figure 4. Ultrasound measurement of optic nerve sheath diameter at 3 mm behind the globe shows thickening and dilatation

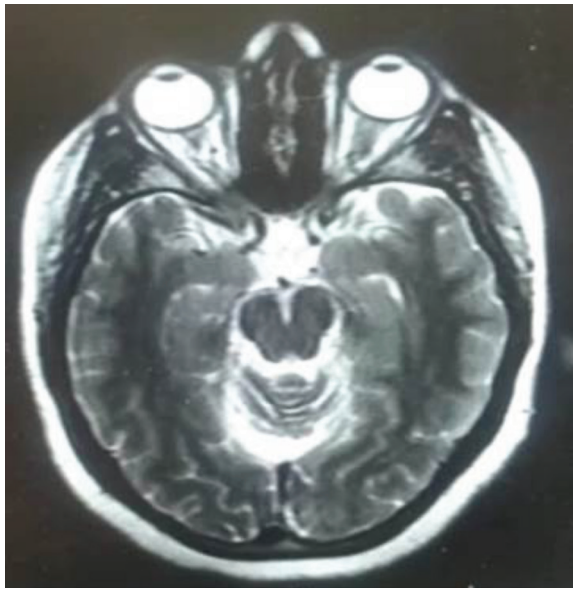

Figure 5. Magnetic resonance imaging of the orbit shows flattening of the left sclera with tortuous left optic nerve and prominent surrounding cerebrospinal fluid

canal was nearly the same in both eyes in cases of bilateral, very asymmetric papilledema using computed tomography, which is more accurate in assessing the bony canal than MRI used by Bidot et al.[16] In our case, the bilaterally distended ONSD makes this theory unsuitable to explain this unilateral condition.

The subarachnoid spaces of the optic nerve are supposed to be inhomogeneous, containing trabeculae, septa, and pillars that might affect CSF dynamics and may have a role in unilateral papilledema.[18] On the other hand, orbital compartmentalization due to decreased communication between the intracranial and intraorbital subarachnoid spaces occurs in IIH and has been implicated as a cause of unilateral papilledema.[19]

Increased collagen in the lamina cribrosa and decreased elasticity, especially with aging, has been suggested as another explanation for unilaterality. Hayreh[20] explained that the optic nerve sheath comprises fibrous tissues that can expand and unfold so that it may expand in both eyes despite only one eye showing higher pressure to cause papilledema. Although many factors may play a role in the unilaterality of papilledema, the most acceptable theory in our case may be orbital compartmentalization. This theory was supported by MRI findings of a tortuous optic nerve on the affected side despite bilateral prominent subarachnoid

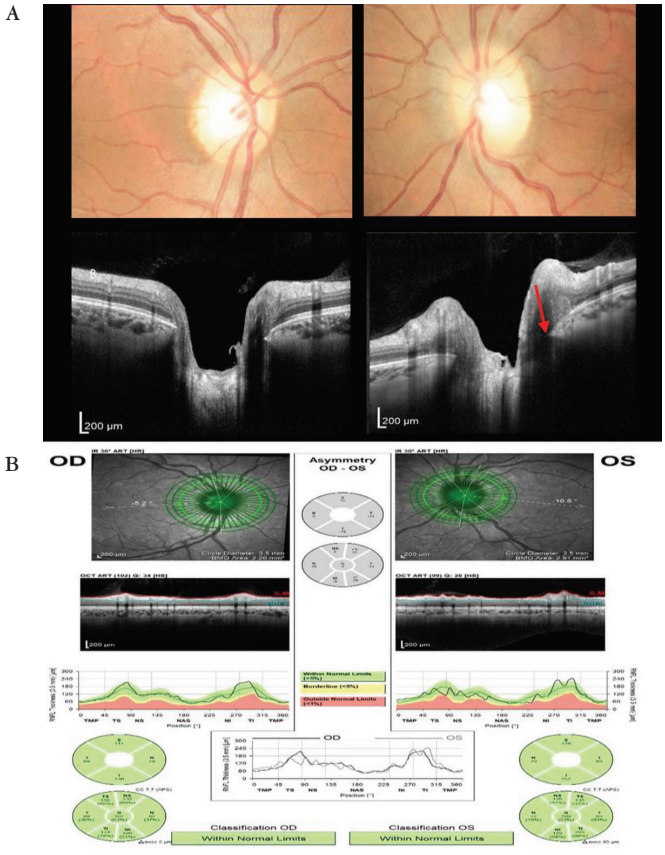

Figure 6. Follow-up images after 4 months. (A) Fundus photography shows improved definition of the disc margins after regression of the previously observed peripapillary fluid. (B) Optical coherence tomography (OCT) line scans of the disc demonstrate peripapillary fluid regression and reduced optic disc height in the left eye. Notice the inferior displacement of Bruch's membrane after resolution (red arrow). The bottom panel shows OCT disc imaging demonstrating a significant reduction in left retinal nerve fiber thickness (RNFLT) and near-normal double hump curve. Right RNFLT appears more or less normal

CSF and increased ONSD on both sides. Another explanation may be an altered response of the optic disc collagen fibers to back pressure or optic nerve head ischemia. Consequently, further studies are required to elucidate these findings.

Unilateral papilledema is a rare condition with unknown pathogenesis, making it challenging to diagnose. However, it should be recognized early to begin prompt treatment to save the optic nerve. Measurement of ONSD may have a role in identifying increased ICP and distinguishing it from other causes of unilateral disc swelling.

Ethics

Informed Consent: Obtained.

Authorship Contributions

Surgical and Medical Practices: R.S.E-G., A.E-S.A.G., Concept: E.M.G., N.A.H., Design: E.M.G., R.S.E-G., Data Collection or Processing: R.S.E-G., A.S.A.E-H., Analysis or Interpretation: E.M.G., A.E-S.A.G., Literature Search: R.S.E-G., Writing: R.S.E-G., E.M.G., N.A.H., A.E-S.A.G.

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