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Case Report

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An unusual case of pseudo foster kennedy syndrome

Abstract

We are hereby presenting a case of Pseudo Foster Kennedy syndrome, which is a rare phenomenon in itself because of normal level of inflammatory markers i.e. ESR and CRP levels with positive temporal artery biopsy.

Keywords: pseudo foster kennedy syndrome, ESR, CRP

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Jatinder Singh Bhalla, Pooja Lal
Department of Ophthalmology, Deen Dayal Upadhyaya Hospital, India

Correspondence: Jatinder Singh Bhalla, Deen Dayal Upadhyaya Hospital, C 79 Chander Nagar, New, Delhi, India, Tel 9718990191, Email jsb_21@yahoo.com

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Abbreviations: AAION, arteritic anterior ischaemic optic neuropathy; ANCA, anti neutrophil cytoplasmic antibody; CRP, C-reactive protein; ESR, erythrocytic sedimentation rate; GCA, giant cell arteritis NA AION, non arteritic anterior ischaemic optic neuropathy; OCT, optical coherence tomography; PFK, pseudo foster kennedy syndrome

Introduction

Foster Kennedy Syndrome (Gowers-Paton-Kennedy syndrome/ Kennedy's Phenomenon/Kennedy's syndrome) - refers to a constellation of findings associated with tumours of the frontal lobe. This syndrome is due to - optic nerve compression, olfactory nerve compression, increased intracranial pressure secondary to a mass (such as meningioma or plasmacytoma usually an olfactory groove meningioma) resulting in optic atrophy in the ipsilateral eye, optic disc edema in the contralateral eye, central scotoma in the ipsilateral eye and ipsilateral anosmia. Pseudo-Foster-Kennedy syndrome is defined as one sided optic atrophy with papilledema in the other eye but with the absence of intracranial mass lesion.

Case report

A 67 year old male presented to our OPD with complaints of dimness of vision in right eye since 12 weeks and dimness of vision in left eye since 3 weeks. Dimness of vision in both eyes was acute in onset, severe and rapidly progressive. It was associated with recurrent mild to moderate global headache which was relieved with analgesics. He was a known smoker, hypertensive (under control with treatment), and non diabetic and non alcoholic. There was no positive history of transient visual obscurations (Amaurosis fugax) or double vision, pain, photopsia, exaggeration of symptoms on heat or exercise, jaw claudication, shoulder pain, head injury, periocular trauma, emotional disturbance, intake of any unknown or toxic substance or medicine, radiation exposure, anosmia, parosmia, fever, weight loss, vomiting, sleep disturbance, no past history of using any eye drop, ocular surgery, or history of similar disease in family. He was being treated in neurology department since last three weeks with complaints of severe headache. Systemic examination was essentially within normal limits except scalp tenderness present on left side temporal area. His left temporal artery was tender with decreased pulsation and right temporal artery was non tender, cord like with absent pulsation. On ocular examination, right eye had visual acuity of counting finger close to face and in left eye was counting finger at 4 metres. Pupillary reactions in both eyes were sluggish to direct and indirect light. Anterior chamber and vitreous showed no cells or flare in both eyes. Right eye disc (Figure 1) was normal in size, with well defined margins, chalky white in colour and loss of optic cup. Left eye disc (Figure 2) was normal in size with ill defined margins, superior sectoral swelling and hyperemic with absence of spontaneous venous pulsations. There were no haemorrhages or exudates on the disc. Visual fields of right eye were severely depressed and left eye showed inferior altitudinal field defect. Fundus Fluorescein angiogram of right eye showed dark appearing optic disc with filling defect and absence of surface capillaries. Left eye angiogram showed delayed filling of choroid and delayed perfusion of the optic disc particularly in the superior sector in the early phase. In the mid phase dilated capillaries were present on the surface of the disc which leaked in the late phase. OCT of optic nerve head and retinal nerve fibre layer showed optic atrophy right eye and optic disc edema in left eye. Haemogram was within normal limits with Hb 12.5 mg/dl and ESR 10 mm/hr. renal function test, serum electrolytes, liver function test, blood sugar levels were within normal limits. Lipid profile was deranged with elevated total serum cholesterol levels 310mg/dl. CSF analysis report was within normal limits. Immunological markers for inflammation like CRP, ANA titre, pANCA, cANCA, VDRL titre, serum ACE level, FTA ABS and serum B_{12} and folic acid levels were normal. Radiological investigations of head and orbit showed no space occupying lesion. Multiple chronic lacunar infarcts and gliotic changes were seen in brain parenchyma in MRI. Bilateral carotid doppler showed presence of non significant haemodynamic plaques. So we arrived at the diagnosis of Pseudo Foster Kennedy Syndrome. The patient was managed by giving IV pulse steroid therapy followed by a tapering dose of oral prednisolone. Following pulse steroid therapy there was no improvement in vision right eye, vision in left eye marginally improved (before treatment counting finger 4 metres and after treatment counting finger 6 metres).

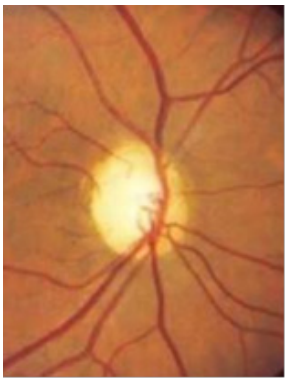

Figure 1 Right eye optic atrophy.

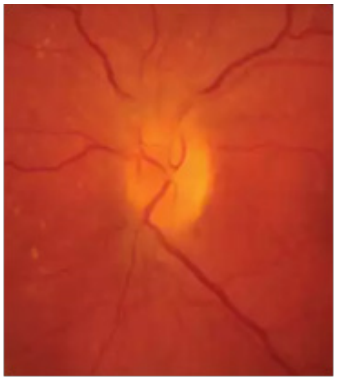

Figure 2 Left eye hyperemic swollen disc.

Discussion

The laboratory hallmark of giant cell arteritis (GCA) is an elevation in the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level. The ESR usually exceeds 50 mm/h and may exceed 100 mm/h, but may be normal in 7-20% of patients with GCA.[1,2] Therefore, a normal ESR does not rule out GCA, and the level of elevation of ESR does not correlate reliably with the severity of the disease. Because normal values of ESR are known to increase with age and are higher in women, the ESR should be adequately adjusted.[3] The CRP is of hepatic origin, usually rises before ESR in most disease states, and is often elevated in GCA. It has higher sensitivity and specificity than ESR (98.6% and 75.7%, respectively) and is relatively unaffected by age, gender, and other hematologic parameters.[4] Non-concordance between ESR and CRP can occur (i.e., either an elevated ESR with normal CRP or a normal ESR with an elevated CRP). The use of both tests provides a slightly greater sensitivity for the diagnosis of GCA (99%) than the use of either test alone.[5]

Giant cell arteritis is diagnosed when clinical suspicion from characteristic clinical symptoms and signs is supported by simple blood tests, including a raised erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), and confirmed by a positive temporal artery biopsy. In occult giant cell arteritis, where there is ocular involvement by giant cell arteritis without any systemic symptoms and signs of giant cell arteritis.[6] the diagnosis is more difficult, and the above investigations will help direct the ophthalmologist before a temporal artery biopsy is carried out. However, it is known that a normal ESR does not preclude the diagnosis of giant cell arteritis.[7] A raised CRP may be a more sensitive indicator of the condition.[8]

A low value of ESR is not very rare in cases of active giant cell arteritis. The reason for such is that the inflammation may be localized or sometimes the immune system is not able to respond by increasing the level of acute phase reactants in body. However, there are well documented reports of cases of biopsy-proven GCA with normal ESRs at diagnosis before glucocorticoid therapy has been started.[9–12] The frequency of such cases is not accurately known. Estimates of the prevalence of a normal pretreatment ESR have varied considerably, and in series of patients with GCA a normal ESR at diagnosis has been reported in 0% to 22.5% of patients.[12–19] Because an elevated ESR is considered a critical marker of GCA, the diagnosis may be delayed when the ESR is normal in a patient who is otherwise suspected of having GCA. In such instances serious vascular complications due to GCA could ensue. Awareness of the actual frequency of normal ESR in GCA should provide the clinician with a perspective that facilitates diagnosis in suspected cases and early implementation of appropriate therapy.

According to literature, systemic symptoms (malaise, fever, or weight loss) are more common in patients with high ESR value. Vision complaints are also more frequent in patients with an elevated ESR.[2] Factors that may influence low level of inflammatory markers in a case of GCA include case selection, previous glucocorticoid treatment, differences in the cutoff value considered to be discriminant for an elevation of the ESR, and the inclusion of patients with "pure" PMR, who may have a lower acute phase response more often than patients with GCA. Two studies of PMR found normal ESR in approximately one fifth of the patients.[20,21] Patients suspected to have GCA with a normal ESR value are often under diagnosed because they are less likely to undergo biopsy. Two recent studies have reported a much higher frequency of normal ESR values in patients with PMR referred to 2 rheumatologic centers.[22,23] A population study (Olmsted County, Minnesota) found 17 of 245 patients with PMR (7.3%) to have an ESR of 40 mm/hour or less at diagnosis.[24]

Conclusion

Thus normal ESR and CRP are not unusual as in our case. Thus in cases of normal ESR and CRP in GCA there should not be delay in starting steroid therapy as it is a neuro ophthalmic emergency. Presence of pale disc in one eye and swollen hyperemic disc in other eye does not always mean Foster Kennedy Syndrome. The occurrence of GCA with normal ESR and CRP is a rare phenomenon. Normal levels of inflammatory markers should not lead to delay in initiating steroid therapy in a sight threatening condition which is a neuro-ophthalmic emergency.

The common causes of Pseudo Foster Kennedy Syndrome are Non Arteritic Anterior Ischaemic Optic Neuropathy (NAAION) and Arteritic Anterior Ischaemic Optic Neuropathy (AAION) with Giant Cell Arteritis. Other less common causes of unilateral disc edema are hypertensive optic neuropathy, retro bulbar neuritis, diabetic papillopathy, pachymeningitis related to microvasculitis, idiopathic intracranial hypertension, unilateral optic nerve hypoplasia and Lebers hereditary optic neuropathy (LHON). AAION has a female preponderance affecting older age group in which visual acuity is severely impaired. There is associated history of headache, scalp tenderness; jaw claudication, proximal muscle weakness and the inflammatory markers i.e. ESR and CRP values are high. Temporal artery biopsy is positive for inflammatory changes in vessel wall.

NAAION usually affects younger age group with vision being mild to moderately impair. It is often associated with sleep apnoea syndrome, nocturnal hypotension, diabetes and hypertension. There is hyperemic swelling of disc with few peripapillary splinter haemorrhages. In hypertensive optic neuropathy there are flame shaped haemorrhages at the disc margin, blurred disc margins, congested retinal veins, papilloedema, and secondary macular exudates. Retrobulbar neuritis usually involves age between 20-50 years. Vision is in range of smelled 6/18 to 6/60 with pain, photopsia and Uhthoff's phenomena. The optic nerve head appears normal. Diabetic papillopathy is usually associated with other diabetic changes in fundus like microaneurysms, dot and blot haemorrhages, cotton wool spots and hard exudates.

Pachymeningitis related to microvasculitis have positive inflammatory markers eg p ANCA, c ANCA with systemic features of microvasculitis. Idiopathic Intracranial Hypertension is related to hypervitaminosis A, usually bilateral, may be unilateral, does not usually result in optic atrophy. LHON (Lebers Hereditary Optic Neuropathy) affects males between age group 15 to 35 years; clinical picture is somewhat similar to AAION except that FFA shows no leakage from the disc or microangiopathic vessles. American college of Rheumatology 1990 classification criteria for GCA states that a patient is said to have GCA if at least three of these five criteria are present. [6]

a. Age at disease onset 50 years or older.

b. New headache.

c. Temporal artery tender on palpation or decreased pulsation.

d. ESR >= 50 mm/hr.

e. Abnormal artery biopsy.

Occult giant cell arteritis has been variously said to occur in 8-38% of cases of giant cell arteritis. [25] In patients with occult giant cell arteritis, both ESR and CRP levels may be significantly lower than in patients with the typical systemic features of giant cell arteritis.[26] The laboratory hallmark of diagnosis of giant cell arteritis (GCA) is an elevation of inflammatory markers like erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level. The combination of the two provides the best specificity (97%). It is well known that lab tests in GCA have decreased specificity, thus clinical picture is important for diagnosis. The ESR may be normal in 7-20% of patients with GCA.[1,2] CRP levels may be normal in 1.7% of patients with GCA. [5] Non-concordance between ESR and CRP can occur (i.e, either an elevated ESR with normal CRP or a normal ESR with an elevated CRP).[5] There are few studies available which state both normal ESR and CRP values in patients with Giant Cell Arteritis at diagnosis.[2,5,27,28] The frequency varies between 4-14% depending on the study and the definition of normal values. Therefore, a normal ESR and CRP level does not rule out GCA. The low acute phase response seems to be related to an intrinsic decreased ability to respond to a phlogistic stimulus. In these patients a genetically determined inhibition in the initiation of the cellular and cytokine cascades involved in the complex process of acute phase response may be hypothesized.[6] Cid and colleagues correlated a low inflammatory response with a high risk of developing visual loss and other cranial ischemic complications. [29–36]

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Conflicts of interest

Author declares that there is no conflict of interest.

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