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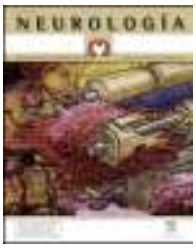

LETTERS TO THE EDITOR

Fluctuating diplopia as an initial manifestation of polycythemia vera☆

Diplopía fluctuante como manifestación inicial de la policitemia vera

Dear Editor:

Fluctuating diplopia as the sole initial manifestation of polycythaemia vera (PV) has never been described in the literature. We present the case of a woman diagnosed with PV who initially presented symptoms of fluctuating diplopia.

Clinical case

Our patient was a 60-year-old woman with a history of chronic migraine, atrial fibrillation, hypercholesterolaemia, and hyperuricaemia. She had undergone mitral valve replacement due to mitral insufficiency and was being treated with anticoagulants. For over a year, she had been experiencing approximately 3 episodes per month of binocular vertical diplopia lasting a few minutes and followed by instability and dizziness. She was simultaneously being examined by the haematology department for polycythaemia, which had been detected 4 months previously in a routine blood test. The general examination was normal; no adenopathies or organomegalies were identified. Baseline oxygen saturation was 99%. The neurological examination revealed left eyelid retraction but was otherwise normal. Eye movement, visual acuity, pupils, eye fundus, and visual field study results were normal. Brain and orbit MRI results were also normal; no pathological contrast enhancement was observed. The routine blood analysis showed a haemoglobin level of 19.1 g/dL, 60.5% haematocrit, 6.51×10^6 red blood cells/μL, a mean corpuscular volume of 92.9 fL, 13.4×10^3 white blood cells/μL, and 309 000 platelets/μL. These results were compatible with a myeloproliferative disorder. Erythropoietin levels were slightly decreased (2.28 mIU/mL); the rest of the iron profile and levels of vitamin B_{12}, folic acid, and alkaline phosphatase were normal. Abdominal ultrasound yielded normal results. A genetic study found a mutation in *JAK2*, which confirmed the diagnosis of PV. The patient underwent therapeutic phlebotomy and began treatment with hydroxycarbamide. In a follow-up consultation 6 months later, the patient showed improved blood test results and reported no new episodes of diplopia or dizziness.

Discussion

PV is a chronic myeloproliferative disorder affecting haematopoietic stem cells. It is characterised by excessive proliferation of red blood cells, white blood cells, and platelets. Criteria for diagnosis include hypercellularity, presence of splenomegaly, and mutation of the *JAK2* gene.[1] It produces a variety of systemic complications with ophthalmologic manifestations, which are normally a consequence of hyperviscosity and thrombosis. They include cerebrovascular accidents, amaurosis fugax, monocular vision loss,[2] anterior ischaemic optic neuropathy,[3,4] retinal haemorrhages, and isolated cranial nerve palsy.[5] Diplopia associated with PV has been linked to ischaemic events occurring in the brainstem at the level of the oculomotor nerve nuclei and pathway or in the nerves themselves. Anatomical differentiation is complex in many cases. In our patient, episodes of fluctuating diplopia were linked to microvascular lesions in the oculomotor nerves as a consequence of hyperviscosity. No further episodes have occurred since treatment was started and blood test results have returned to normal, meaning that the predisposing factor has been removed. Diplopia in the context of PV is rare. Ours is the first case of fluctuating diplopia reported in the literature. In-depth examination and cooperation with haematologists is essential in patients with diplopia in order to determine the appropriate treatment and avoid new ischaemic events.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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Intrathecal baclofen for dystonia treatment during pregnancy: A case report☆

Baclofeno intratecal para el tratamiento de la distonía durante el embarazo: un caso clínico

Dear Editor:

Baclofen is an anti-spasmodic drug and muscle relaxant indicated for treating spasticity.[1,2] The exact mechanism of action is unclear, although it is believed to be a GABA agonist; it decreases the amount of free aspartate and glutamate, thereby reducing the excitability of alpha motor neurons.[3] Typically, baclofen is administered orally to treat spasticity produced by certain conditions. For cases which do not respond adequately to oral administration, or which come with intolerable side effects, there are continuous infusion pumps that administer baclofen intrathecally.[4] Baclofen has also been used to treat some cases of dystonia, whether focal or generalised, in patients who do not respond to conventional oral treatments or botulinum toxin injection.[5,6] To date, the effects of baclofen use during pregnancy have yet to be fully examined. The FDA has identified baclofen as a category C risk for pregnant women: 'Animal studies have revealed adverse fetal effects but no controlled studies in women have been done, or no studies in women or animals are available.' In animal studies completed thus far, baclofen was administered orally and at high doses. No prospective controlled studies evaluating the safety of intrathecal delivery of this drug during pregnancy have been carried out. Furthermore, this medication is prescribed only infrequently in clinical practice.[7] The purpose of our letter is to provide additional information about baclofen use during pregnancy by presenting a clinical case.

The case is that of a 30-year-old woman with autosomal dominant DYT1-positive generalised dystonia, with symptoms beginning when she was 11. After various oral medications and an injection of botulinum toxin type A elicited no response, doctors opted to implant an intrathecal baclofen pump at the lumbar level (SynchroMed® II 40 ml, as she had responded to a test dose of 100 μg). After implantation, the patient remained essentially asymptomatic for 3 years, at which point she became pregnant. She was informed of the possible risks and disadvantages, and decided with her doctors not to have the pump removed, given the good response up to that point. She therefore received a continuous infusion of baclofen dosed at 200 μg/day (8.3 μg/h) during her entire pregnancy. The patient's pregnancy progressed uneventfully and she gave birth at week 39 to a healthy 3080 g baby girl through vacuum-assisted vaginal delivery, without an epidural or any other complications. The patient reported no increases in dystonia symptoms either during or after her pregnancy. The patient breastfed for the first month after which she stopped for personal reasons. Four years later, the patient had an ectopic pregnancy which required surgery and the removal of a fallopian tube. Later, her dystonia symptoms worsened, and her medication was increased to control them (current dose, 600 μg/d). The patient remains essentially asymptomatic and is planning another pregnancy. Her daughter is now a healthy 7-year-old with psychomotor development within normal limits.

Articles addressing the effect of baclofen during pregnancy are scarce. Our literature search revealed only 10 reported cases, and there were no prospective controlled studies.[7—14] In all of them, the treatment was indicated for a diagnosis of spasticity secondary to a variety of illnesses, such as cerebral palsy (5 cases),[7,9,13,14] spinal cord injury (3 cases),[7,8,11] tetanus (1 case),[10] and multiple sclerosis (1 case).[12] Nearly all the patients had their pumps implanted between 15 months and 6.5 years before pregnancy, although in 2 cases the pumps were implanted during the third trimester (week 28-30) due to an increase in spasticity. Most of them underwent planned early caesarian

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