often appears in both IIH and in CSF fistulas. Typically, the headache associated with low CSF pressure worsens within 15min after sitting or standing and is often accompanied by neck stiffness, tinnitus, hearing loss, photophobia and nausea.[8] In our patient, we suspected that the headache was due to liquoral hypotension, in addition to liquorrhea, because the pain and dizziness worsened with standing. Lumbar puncture was not performed for the initial diagnostic suspicion, as it might have worsened the headache from intracranial hypotension.

In conclusion, IIH requires close monitoring because complications can occur. A CSF fistula, although rare, is an adverse event that requires early diagnosis and surgical treatment to prevent the development of meningitis or pneumoencephaly.[9]

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Corectopia as a neuro-ophtalmological sign of polyradiculopathy

Corectopia como manifestación neurooftálmica en un caso de polirradiculopatía

Dear Editor:

Corectopia is the manifestation of an eccentric, oval pupil. It is primarily described in mesencephalic pretectal lesions.[1] This alteration is the result of an incomplete injury of the pupillary fibres, which produces a selective inhibition of the iris sphincter tone.[1] We present the case of a patient with inflammatory demyelinating polyradiculopathy who presented corectopia.

The patient was a 32-year-old Caucasian male, with no relevant medical history, who attended the emergency service due to back pain of 15 days' evolution. He also reported intense pain in both sural regions and paresthesia in the soles of the feet and palms of the hands. The neuro-ophthalmic exploration revealed corectopia and anisocoria (right 3mm, left 2mm with ambient light; right 5 mm, left 4 mm with light deprivation) (fig. 1), both with photomotor reflex and accommodation, as well as a slight left peripheral facial palsy. The neurological examination revealed hyporeflexia (+/4+) in the lower extremities and hypoesthesia and allodynia in the soles and palms. Analytical studies, autoimmune profile and vitamins were normal. Serological testing was positive for cytomegalovirus. An electrocardiogram, chest radiograph and cranial and spinal and cranial magnetic resonance imaging showed no abnormalities. A cerebrospinal fluid (CSF) analysis showed mononuclear pleocytosis (70 cells; 90% lymphocytes; elevated protein, 80mg/dl) with normal CSF glucose. The electromyographic analysis showed a predominantly demyelinating sensorimotor polyradiculoneuritis. During his latest check-ups, the patient presented an obvious clinical improvement with partial recovery from the pupillary disorder.

Figure 1 Image showing a corectopia.

Polyradiculitis may affect the autonomic nervous system and be accompanied by pupil abnormalities. Horner syndrome has been described from sympathetic affectation, cranial neuropathies of the 3rd cranial nerve and Adie tonic pupil.[2,3] Our patient presented corectopia within a case of polyradiculitis; the photomotor reflex and lack of a tonic reaction exclude the possibility of an Adie tonic pupil.[4,5] The clinical case with an eccentric pupil is compatible with corectopia. This finding has classically been associated with pretectal involvement[1]; however, it should be regarded as a finding of other processes that may affect the autonomic system. Our case indicates that the most reasonable pathophysiology of corectopia is a lesion of the autonomic fascicle of the 3rd cranial nerve in the midbrain or a peripheral lesion rather than a nuclear lesion.

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Congenital arthrogryposis multiplex and gastroschisis in the same patient

Artrogriposis múltiple congénita y gastrosquisis en un mismo paciente

Dear Editor:

Arthrogryposis multiplex congenita (AMC) is a rheumatic disorder characterised by multiple joint contractures that affect the back muscles as well as the upper and lower extremities. The muscles may be absent, reduced in size and/or number, with replacement by fibrous tissue or fat.[1] The degree of involvement varies from patient to patient and the classical deformity is bilateral and symmetrical.[2] Its prevalence is estimated at 1/3,000 newborns, without distinction by race or gender, and a genetic cause is found in only 30%of cases.[2]

The aetiology of this condition remains unknown and many causal agents have been implicated, particularly viral infections and intrauterine restrictive disorders. The possibility of a vascular rupture phenomenon has also been mentioned as a possible cause.[3,4]

Gastroschisis is a congenital defect of the anterior abdominal wall, lateral to the umbilical cord insertion, usually on the right side. Its aetiology is considered multifactorial and maternal factors such as exposure to teratogens, salicylates and nutritional deficiencies in the early weeks of gestation have currently been mentioned. Any of the previously mentioned teratogenic factors probably causes a vascular alteration of the omphalomesenteric artery (primary defect), which destroys a portion of the abdominal wall, through which abdominal contents protrude into the amniotic cavity.[5-7]

There have been very few published reports of the association between AMC and gastroschisis.[8]

We present a male patient with AMC and gastroschisis, son of a mother aged 36, with 4 pregnancies, who consulted at 36 weeks of gestation to receive delivery care. Weight at birth was 2,400g; head circumference was 29cm; size was 43cm. The mother followed no prenatal care and underwent no obstetrical ultrasound. She denied consumption of tobacco, alcohol or psychoactive substances. The patient was assessed by the paediatric neurology service, which requested brain scans. These were reported as normal and paediatric surgery was carried out to correct the abdominal wall defect, which was corrected on the second day of life. Karyotyping with G "banding" was requested; it reported a normal chromosome complement (46XY). Echocardiography was also requested and was reported as normal.

The association of gastroschisis with other birth defects with a possible aetiology of vascular disruption, such as Poland sequence, intestinal atresia and AMC, has been reported.[8] A recent publication aimed to assess the frequency and type of malformations associated with gastroschisis in 24 different registries of birth defects that contributed data to the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR); this publication collected information from 3,322 patients with gastroschisis, of which 17 also presented AMC. Although the aetiology of these two conditions is not clear, it has been noted that a vascular disruption phenomenon may be involved. This phenomenon may explain the coexistence of these two conditions in our patient.[9]

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