A RARE CAUSE OF UNILATERAL HEADACHE AND OPHTALMOPLEGIA

UNE CAUSE RARE DE CÉPHALÉE UNILATÉRALE ET D'OPHTALMOPLÉGIE

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Abstract

Tolosa-Hunt syndrome is a painful ophthalmoplegia, usually unilateral, caused by nonspecific inflammation of the cavernous sinus or superior orbital fissure. It is a rare entity of unknown etiopathogenesis. We report the case of a 26-year-old woman who presented with a left headache, diplopia and painful eye movements. Examination revealed left ptosis, exophthalmos and ophthalmoplegia. Brain magnetic resonance imaging revealed left orbital apexitis. The immunological and infectious assessments were negative. The patient was treated successfully with steroids and remained stable after two years of follow-up. The diagnosis of Tolosa Hunt Syndrome was retained in the face of clinical presentation, normal investigations, magnetic resonance imaging findings and response to steroid therapy. Although it is often a benign condition, it may affect the optic nerve and lead to blindness. Therefore, it is important to recognize this disease and provide adequate treatment to preserve visual function.

Key – Words : Tolosa Hunt Syndrome ; Ophthalmoplegia ; Apexitis

Résumé

Le syndrome de Tolosa-Hunt est une ophtalmoplégie douloureuse, généralement unilatérale, causée par une inflammation non spécifique du sinus caverneux ou de la fissure orbitaire supérieure. C'est une entité rare d'étiopathogénie inconnue. Nous rapportons le cas d'une femme de 26 ans qui s'est présentée avec une céphalée gauche, une diplopie et des mouvements oculaires douloureux. L'examen a révélé un ptosis gauche, une exophtalmie et une ophtalmoplégie. L'imagerie par résonance magnétique cérébrale a révélé une apexite orbitaire gauche. Les bilans immunologique et infectieux étaient négatifs. La patiente a été traitée avec succès avec des stéroïdes et est restée stable après deux ans de suivi. Le diagnostic de syndrome de Tolosa Hunt a été retenu face à la présentation clinique, aux examens normaux, aux résultats de l'imagerie par résonance magnétique et à la réponse à la corticothérapie. Bien qu'il s'agisse souvent d'une affection bénigne, elle peut affecter le nerf optique et entraîner la cécité. Par conséquent, il est important de reconnaître cette maladie et de fournir un traitement adéquat pour préserver la fonction visuelle.

Mots - clés : Syndrome de Tolosa Hunt ; Ophtalmoplégie ; Apexite

ملخص

متلازمة تولوسا هانت هي شلل مؤلم في العين عادة ما يكون من جانب واحد، وينتج عن التهاب غير محدد في الجيب الكهفي أو الشق المداري العلوي. إنه مرض نادر ومسبباته غير معروفة. أبلغنا عن حالة امرأة تبلغ من العمر 26 عامًا تعاني من صداع في الجهة اليسار وشفع وحركات مؤلمة في العين. كشف الفحص عن تدلي الجفن الأيسر وجحوظ العين وشلل العين. كشف التصوير بالرنين المغناطيسي للدماغ عن التهاب قمة الحجاج الأيسر. كانت التقييمات المناعية والمعدية سلبية. تم علاج المريضة بنجاح باستخدام الكورتيكوستيرويد وظلت مستقرَّة بعد عامين من المتابعة. تم الاحتفاظ بتشخيص متلازمة تولوسا هانت أمام العرض السريري، نتائج التقييمات العادية، نتائج التصوير بالرنين المغناطيسي والاستجابة للعلاج بالستيرويد. على الرغم من أنها حالة حميدة في كثير من الأحيان، إلا أنها قد تؤثر على العصب البصري وتؤدي إلى العمى. لذلك، من المهم التعرف على هذا المرض وتقديم العلاج المناسب للحفاظ على الوظيفة البصرية.

الكلمات المفاتيح : متلازمة تولوسا هانت ; شلل العين ;التهاب القمة المدارية.

INTRODUCTION

Tolosa Hunt syndrome (THS) is an inflammatory process of the cavernous sinus, the superior orbital fissure and/or the orbital apex of unknown etiopathogenesis [1]. It is one of the rare disorders recognized by the National Organization for Rare Disorders (NORD) and it is included by the International Headache Society (IHS) in its headache classification as one of the painful cranial neuropathies [2]. The estimated annual incidence is one case per million per year [3]. It presents with unilateral orbital/periorbital pain associated with paralysis of one or more of the IIIrd, IVth or VIth cranial nerves [3]. THS is a diagnosis of exclusion and it responds well to steroids [2]. We report, here, a rare case of THS in a young woman revealed by painful ophthalmoplegia.

CASE PRESENTATION

A 26-year-old healthy female patient presented with complaints of blurred vision, headache, and left eye pain with diplopia. Examination revealed left ptosis with exophthalmos and ophthalmoplegia (figures 1 and 2).

Visual acuity was 7/10 in the left eye and 10/10 in the right eye. Fundoscopic examination was normal in both eyes. The neurologic examination was unremarkable. Laboratory investigations including complete blood count, erythrocyte sedimentation rate, C-reactive protein, serum protein electrophoresis, fasting blood sugar, renal and liver functions, angiotensin-converting enzyme, antinuclear antibodies, antineutrophil cytoplasmic antibodies and viral serologies testing were either normal or negative. The Intradermal tuberculin test was negative. Brain magnetic resonance imaging (MRI) showed left orbital apexitis with hyper intensity on the T2 sequence and contrast enhancement of the left orbital apex extended to the anterior part of the cavernous sinus and oculomotor muscles (figure 3). The patient was treated by intravenous steroids (methylprednisolone 1mg/kg daily) for three days relayed by a high dose of oral steroid (prednisone 2 mg/kg daily) for one month followed by a progressive degression. The Outcome was favorable with a significant reduction of pain in 72 hours and an improvement of oculomotricity and other symptoms within the first week. After two years of follow-up, the patient remained stable without any recurrence.

Figure 1: Left exophtalmos with upper lid ptosis

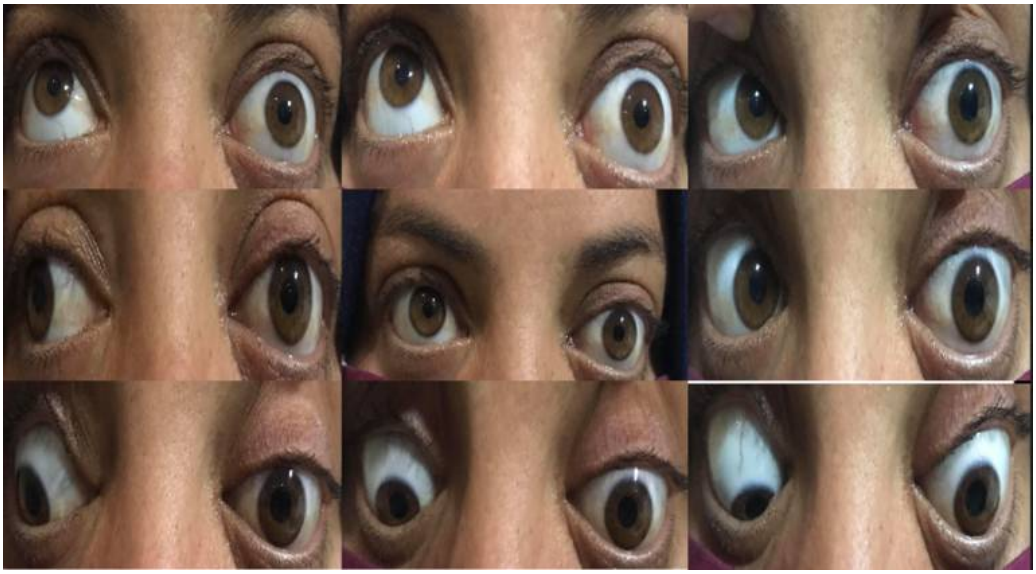

Figure 2: Left ophtalmoplegia with exophthalmos

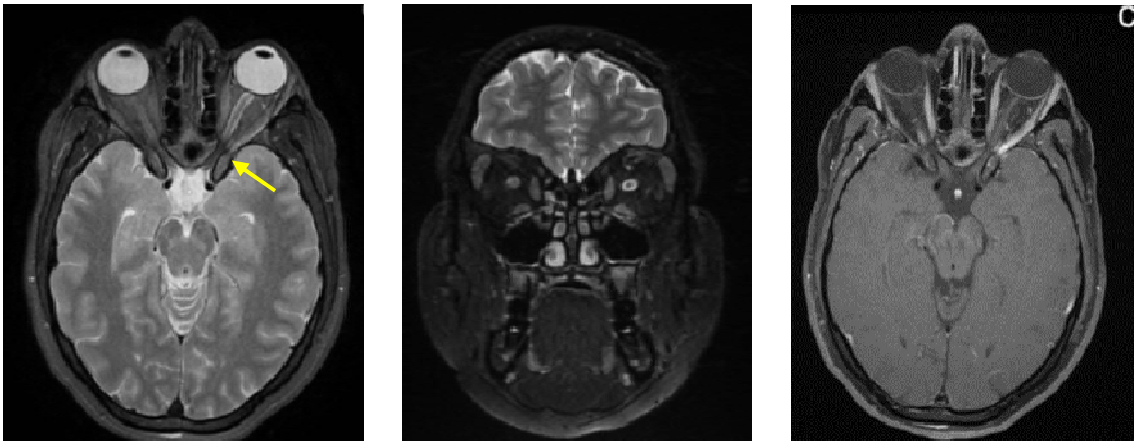

Figure 3: Brain MRI revealing left orbital apexitis with diffuse enhancement of the left orbital apex (yellow arrow) extending to the anterior cavernous sinus and oculomotor muscles.

DISCUSSION

Our case summarizes a rare entity. The diagnosis of THS was retained in the face of painful ophthalmoplegia, the negativity of all investigations carried out as well as spectacular response to treatment and patient stability after two years of follow up. THS, also known as painful ophthalmoplegia, recurrent ophthalmoplegia, or ophthalmoplegia syndrome, is manifested as unilateral periorbital headaches associated with painful and restricted eye movements caused by paralysis of one or more of the IIIrd, IVth and/or VIth cranial nerves [3,4]. It does not have any age or sex predilection [2]. Pathophysiological and etiopathogenic mechanisms of THS remain obscure [5]. It is caused by nonspecific granulomatous inflammation characterized by infiltration of lymphocytes and plasma cells in and around the cavernous sinus, with variable extension into the superior orbital fissure/ orbital apex [3]. The cavernous sinus is the most common site of multiple cranial nerve involvements. Rarely, inflammation can involve the orbital apex leading to optic nerve damage and consequent optic disc pallor or swelling [1]. Loss of visual acuity in those cases is unpredictable and can be permanent. Oculomotor palsy can knock or succumb to orbital pain. All three oculomotor nerves can be affected with different combinations described. Involvement of the common oculomotor nerve, the abducens nerve and the pathetic nerve affects 80, 70 and 29% of cases, respectively [6]. Brain MRI with contrast, especially the coronal view, is a crucial diagnostic

tool and helps to exclude other etiologies [2]. MRI findings of THS show inflammatory changes within the anterior cavernous sinus, with or without involvement of the internal carotid artery, superior orbital fissure, orbital apex, and/or optic nerve. On the affected regions, T1 images may appear isointense with hyperintensity on T2 images and contrast enhancement [7]. THS remains a diagnosis of exclusion. The International Classification of Headache Disorders, 3rd edition (ICHD-3) criteria for THS include a unilateral headache, granulomatous inflammation of the cavernous sinus, superior orbital fissure or orbit, demonstrated by MRI or biopsy, with paresis of one or more of the IIIrd, IVth and/or VIth ipsilateral cranial nerves, a causal relationship of the inflammation with the headache and exclusion of other diagnoses [8].

It was suggested that ICHD-3 criteria are suboptimal for the accurate diagnosis of THS. False-negative cases are not harmful to the patients, as their symptoms remit with the use of steroids. False-positive diagnoses are the most dangerous for patients presenting with symptoms concerning THS. Neurinomas, lymphomas, sarcoidosis, meningiomas, and pituitary macroadenomas may demonstrate radiological features comparable to those of THS. A close follow-up should be performed before a definitive diagnosis of THS is reached. The ICHD-3 criteria were applicable in our patient and she remained stable after two years of follow-up. Considering its inflammatory nature, THS is usually treated with steroids, but there are no rules regarding therapeutic protocol [9,10]. Steroid treatment is given orally or by intravenous injection [10]. It provides rapid resolution of the orbital pain within few days, which also serves as diagnostic confirmation. However, the resolution of neuropathies necessitates a longer course of steroids [9,10]. Our patient had a spectacular response to steroids with a significant reduction of pain in 72 hours and an improvement of oculomotricity within the first week. THS patients have a good prognosis but it is important to consider follow-up as recurrences occur in about 21–50 % of the cases over for months to years [10].

CONCLUSION

Although THS is often a benign condition, it is important to recognize its symptoms and clinical features and to provide immediate treatment to preserve visual and ocular motility functions and to prevent sequelae at a later stage.

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