Simultaneous Bilateral Painful Ophthalmoplegia and Exudative Retinal Detachment in Rheumatoid Arthritis

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Abstract

A 47-year-old woman who suffered from chronic rheumatoid arthritis presented with bilateral painful ophthalmoplegia with proptosis and exudative retinal detachment. The painful ophthalmoplegia with proptosis might have been caused by pachymeningitis involving the cavernous sinus bilaterally, or bilateral posterior scleritis spreading to the extraocular muscles and tendons. The exudative retinal detachment might have been a result of bilateral posterior scleritis which had spread to the choroid. These two unusual complications of rheumatoid arthritis occurred simultaneously in this case. Both complications responded to corticosteroid treatment.

Key word : Painful Ophthalmoplegia, Retinal Detachment, Pachymeningitis, Posterior Scleritis, Rheumatoid Arthritis

Neuro-ophthalmologic complications in rheumatoid arthritis are uncommon, and most are due to rheumatoid nodules which involve the nervous system and meninges[1-4]. The common clinical manifestation of these rheumatoid nodules is ophthalmoplegia[3]. Other sporadic reports such as tenosynovitis of the superior oblique tendon sheath, anterior ischemic optic neuropathy and multiple cranial nerve palsies, are due to inflammation,[5] vasculitis[6] and pachymeningitis,[7] respectively. Pachymeningitis and exudative retinal detachment in rheumatoid arthritis are usually associated with long-standing seropositive, erosive articular disease and are usually accompanied by other extra-articular features[1,2,8-11]. We describe an extremely rare presentation of simultaneous pachymeningitis and bilateral exudative retinal detachment without other extra-articular features in a case of rheumatoid arthritis.

CASE REPORT

A 47-year-old woman was admitted to Songklanagarind Hospital because of bilateral proptosis, chemosis and gradual loss of vision for one week. She had a 9-year history of seropositive multiple joint deformity rheumatoid arthritis without any other extra-articular manifestations. The rheumatoid factor titer was 1:320 in 1994. She received

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aspirin (300 mg daily) and predisolone (5 mg daily) as maintenance therapy. Three months before admission, she had bilateral anterior uveitis which was successfully treated with a topical steroid. Two weeks before admission, she had fever, bilateral ocular pain at rest, aggravated by ocular movement. Her vision gradually decreased. Physical examination revealed a body temperature of 38°C, pulse rate of 100 beats/min, respiratory rate of 20 breaths/min and blood pressure of 100/60 mmHg. Rheumatologic examination disclosed deformities of both hands characterized by ulnar deviation, zigzag, swan neck and boutonniere deformities of the fingers. No active arthritis and other extra-articular manifestations were detected. Neuro-ophthalmologic examination revealed bilateral proptosis and chemosis, as well as bilateral total external ophthalmoplegia. The pupils were 3 mm bilaterally and reactive to light. Bilateral exudative retinal detachment at the macular region was also observed. No inflammation was found in the uveal tract, optic disc and retinal vessels.

During hospitalization, a complete blood count revealed : hemoglobin 6.0 mmol/l (9.7 g/dl), hematocrit 0.26 (26%) and leukocyte 12 $x10^9$ /l (12,000 /mm^3) with 91 per cent Neutrophils, 6.0 per cent Lymphocytes, and 3.0 per cent Eosinophils. Platelet count was $527x10^9$ /l (527,000/mm^3). Rheumatoid factor titer was 1:320 and the erythrocyte sedimentation rate was 66 mm/hour. Cerebrospinal fluid analysis showed acellular CSF with normal CSF pressure as well as normal CSF/plasma glucose ratio and protein. CSF cultures were negative for TB, fungus and other microorganisms. Blood and CSF serology for cryptococcal antigen and syphilis were negative. Urinalysis revealed pyuria and a urine culture was positive for *Proteus Mirabilis*. CT scan and MRI of the brain showed bilateral and symmetrical enhancement of the globe and retinal detachment without enhancement of the extraocular muscles and tendons. These imaging studies also revealed diffuse dural enhancement with extension to sella and parasella regions (Fig. 1 and 2). Dural biopsy revealed fibrous necrotic dura with lymphocyte infiltration (Fig. 3). No classical rheumatoid nodule or vasculitis was detected. Tissue stain and culture for acid fast bacilli, bacteria and fungus were negative. Ceftazidime (6 g daily) for urinary tract infection and prednisolone (50 mg daily) for active extra-articular involvement from rheumatoid arthritis were employed. The neuro-ophthalmologic abnormalities gradually improved. Her ocular movement was normal within one month and her visual acuity became 20/30-1 OD and 20/70 OS within two months. The retinal detachment disappeared in 3 weeks.

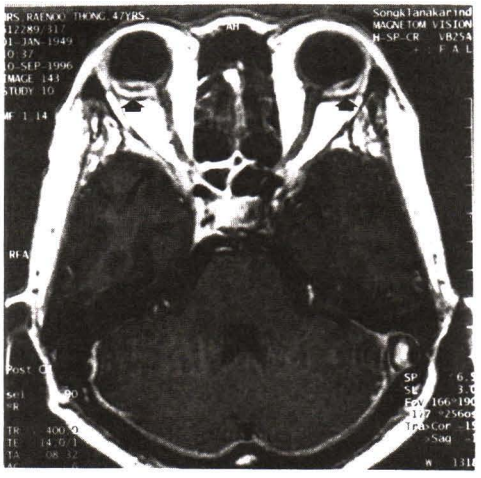

Fig. 1. Axial T1-weighted gadolinium-enhanced MRI showing diffuse enhanced meninges with extension to sella and parasella regions (curved arrows). Bilateral enhanced globes with retinal detachment (straight arrows).

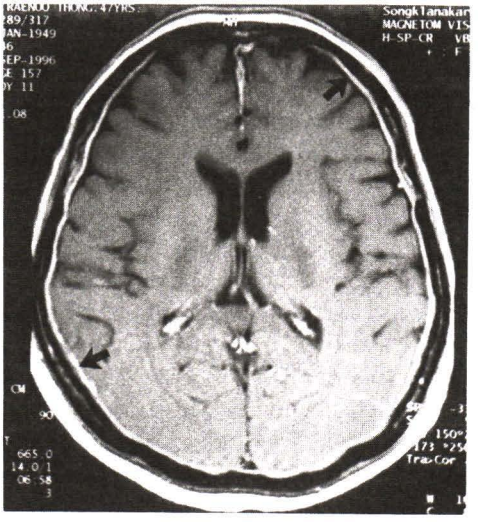

Fig. 2. Axial T1-weighted gadolinium-enhanced MRI showing diffuse enhanced meninges (straight arrows).

Fig. 3. Microscopic picture of dural biopsy showing fibrous, necrotic dura with lymphocyte infiltration without vasculitis (hematoxylin-eosin, original manification x 40).

DISCUSSION

Rheumatoid pachymeningitis, an extremely rare complication of rheumatoid arthritis is characterized by focal or diffuse fibrous thickening of the dura[1,7,8,12]. The microscopic picture shows variable findings which include fibrosis with or without inflammatory cell infiltration such as lymphocyte, plasma cell and multinucleated giant cell[1,7,8,12]. Rheumatoid nodules are frequently detected in the affected dura[1-4] and necrotic debris is occasionally observed[12,13]. The pachymeningitis may be asymptomatic or may be associated with headache and multiple cranial nerve palsies[1,7,8,10]. Spinal cord or nerve root compression have rarely been reported[12,13].

Exudative retinal detachment is also a rare complication of rheumatoid arthritis[14,15]. The mechanism of retinal detachment is due to inflammation of the choroid or inflammation spreading from an adjacent area[14,15]. The clinical presentations usually consist of bilateral or unilateral decrease of visual acuity and ocular pain[14,15]. CT and MRI scan of our case showed scleral thickening with enhancement which was compatible with posterior scleritis[16]. Posterior scleritis is also a rare complication and occurs in 0.62-6.3 per cent of rheumatoid arthritis[11,15]. The most common features are orbital pain, decreased vision and redness of the eye[10,15]. Proptosis, ptosis and diplopia may be seen in some cases due to inflammation spreading to the extraocular muscles and tendons[11]. The imaging studies may show scleritis with concomitant orbital pseudotumor or myositis[16]. Although fluorescein fundus angiography was not done in our case, we believed that exudative retinal detachment was an inflammatory extension of posterior scleritis to the choroid.

The pachymeningitis in our case was diffuse. However, the clinical presentation of bilateral external ophthalmoplegia and chemosis might indicate prominent involvement of the dura around the sella and parasella regions. It might be the extension of bilateral posterior scleritis to extraocular muscles and tendons but enhanced CT and MRI showed no evidence of myositis or orbital pseudotumor.

Rheumatoid pachymeningitis and exudative retinal detachment with posterior scleritis are associated with long-standing seropositive, erosive arthritis and usually accompanied by other extra-articular manifestations such as subcutaneous nodules, pulmonary nodules and pericardial effusion[1,2,8-11]. Our patient had diffuse pachymeningitis and bilateral retinal detachment with posterior scleritis without previous extra-articular manifestations. She also had a long history of rheumatoid arthritis with no previous extra-articular manifestations. This is unique and may be the first report of such an extra-articular presentation.

The clinical course of rheumatoid pachymeningitis is subacute or chronic, lasting from weeks to months or years[1,7,10,12]. Treatment consists of corticosteroid, cytotoxic agents and/or surgical decompression[1,4,12,13]. The prognosis of pachymeningitis varies; but is usually not good [1,8]. This may be explained by the progressive fibrotic process found in most pathologic studies. Unlike pachymeningitis, exudative retinal detachment with posterior scleritis usually responds to corticosteroid therapy. However, cytotoxic agents, penicillamine or gold may be advocated in some cases[9,11,15].

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ภาวะปวดตาสองข้างกลอกตาไม่ได้และจอประสาทตาลอกที่เกิดขึ้นพร้อมกันในผู้ป่วยข้ออักเสบรูมาตอยด์

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ผู้ป่วยหญิงอายุ 47 ปี เป็นโรคข้ออักเสบรูมาตอยด์เรื้อรัง มาด้วยอาการตาทั้งสองข้างปวด กลอกตาไม่ได้ ร่วมกับตาโปน และจอประสาทตาลอก อาการปวดตากลอกตาไม่ได้ร่วมกับตาโปน อาจเกิดจากเยื่อหุ้มสมองอักเสบหนาตัวลุกลามเข้าไปในคาเวอร์นัสไซนัสทั้ง 2 ข้าง หรือการอักเสบของตาขาวด้านหลังทั้งสองข้างลุกลามไปยังกล้ามเนื้อ และเอ็นของตา จอประสาทตาลอกอาจเป็นผลจากการอักเสบของตาขาวด้านหลังทั้งสองข้างลุกลามไปถึงคอรอยด์ ภาวะแทรกซ้อนทั้งสองของโรคข้ออักเสบรูมาตอยด์พบได้ไม่บ่อย ในผู้ป่วยรายนี้ภาวะแทรกซ้อนทั้งสองเกิดขึ้นพร้อมกัน และตอบสนองต่อการรักษาด้วยคอร์ติโคสเตียรอยด์

คำสำคัญ : ปวดตา, กลอกตาไม่ได้, จอประสาทตาลอก, เยื่อหุ้มสมองอักเสบหนาตัว, ตาขาวด้านหลังอักเสบ, โรคข้อรูมาตอยด์

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