Unilateral Facial Swelling and Exophthalmos in a Patient with Polyarteritis Nodosa

Kazuyuki Ishida, Tsukasa Yokota, Yoshiaki Wada, Masahito Yamada and Hiroshi Tsukagoshi

A patient with polyarteritis nodosa (PN) presenting with exophthalmos and facial swelling, which are rarely found in PN, is reported. The patient, a 27-year-old male, complained of painful facial swelling and diplopia. Physical examinations showed facial swelling around the right orbit and exophthalmos. After admission, he experienced myalgia in both calves. Laboratory studies disclosed leukocytosis and liver dysfunction. Celiac and renal angiograms showed aneurysms. A biopsy specimen of his left calf showed an arterial inflammatory process. The patient was diagnosed as having PN. He had an excellent response to corticosteroids, with prompt improvement.

(Key words: ocular proptosis, systemic vasculitis, MRI scan, muscle biopsy, steroid therapy)

Introduction

Polyarteritis nodosa (PN) is a systemic disease involving small- and medium-sized arteries and veins of many organ systems, including the kidney, heart, gastrointestinal tract, muscle and nervous system, with widely varied manifestations (1). Morbidity and mortality are related to the type and degree of organ system involvement (1).

The eye, orbit, and skin are affected in about 10% of patients with PN, as types of systemic vasculitis or as local, atypical forms (2, 3). Common ophthalmologic complications of PN are episcleritis, hypertensive retinopathy, central retinal artery occlusion, choroidal insufficiency, extraocular muscle dysfunction and ischemic optic neuropathy (2, 4, 5). Cutaneous involvement of PN is also common, which includes ulcers, livedo, nodules and gangrene (6). However exophthalmos and facial swelling secondary to PN are very rare (5, 7–11). Such symptoms are commonly found in other connective tissue diseases like Wegener’s granulomatosis (12–14).

Here, we report a case of PN presenting with exophthalmos and facial swelling.

Case Report

In January 1990, a 27-year-old man, an engineer, was referred from an oral surgeon to our neurologic clinic. He complained of painful swelling of the right face and double vision. The patient had been in good health until one month prior to admission to our hospital, when he noticed painful unilateral facial swelling around the right eye, followed by a low grade fever and diplopia. He underwent examination at a dental hospital, was diagnosed as having Quincke’s edema, and was treated with oral prednisolone (30mg daily), resulting in improvement of his symptoms. However, three weeks later the same symptoms recurred, with additional symptoms of impaired vision of the right eye, lower abdominal pain, and myalgia in both calves.

At our initial evaluation in January, he had a remittent fever up to 40°C, his blood pressure was 134/90mmHg, and pulse rate was 88/min. There was an obvious swelling on the right side of the face around the orbit with a puffy eyelid, a narrow palpebral fissure, slight proptosis, and an injected conjunctiva on the right side (Fig. 1, left). There were no abnormalities in the oral or nasal cavity. Lymphadenopathy was absent. The chest was clear to percussion and auscultation, and palpation of the abdomen was negative. Nodules, skin rash, and edema were not found. Neurological examination revealed diplopia in all directions. The ocular movements were within normal limits. Visual acuity was 0.2 on the right and 0.5 on the left. Visual fields were intact. Funduscopic
examination revealed no papilledema, retinal hemorrhage, or edema. The patient had muscle pain in both calves. There was no weakness of muscles, except for the plantar flexors of both ankles. The myalgia prevented him from plantar flexion of the ankles. The deep tendon reflexes of the extremities were equal and normoactive, and no abnormal reflexes were found. Sensory abnormalities were not noted.

Laboratory findings on admission were as follows: the white blood cell count was 19,200/mm³, with 77% neutrophils and 8% eosinophils. The platelet count was 19.2 x 10⁴/mm³. Serum electrolyte values were all within normal limits. The urea nitrogen was 11 mg/dl, the creatinine 0.9 mg/dl. Fasting plasma glucose concentration was 145 mg/dl. Liver function tests showed total bilirubin 0.9 mg/dl, lactic dehydrogenase 364 U/l, glutamic oxaloacetic transaminase 38 U/l, glutamic pyruvic transaminase (GPT) 122 U/l, alkaline phosphatase 582 U/l, and gamma-glutamyl transpeptidase 237 U/l. The creatine kinase level was normal and the amylase value was slightly high. The erythrocyte sedimentation rate was 78 mm/h. C-reactive protein (CRP) was 6+. Tests for rheumatoid factor, antinuclear antibody, and circulating immune complexes were negative. A quantitative analysis of immunoglobulins disclosed that IgG was 616 mg/dl, IgM below 46 mg/dl, and IgA 167 mg/dl. The serum C₃ level was 154 mg/dl, C₄ 51 mg/dl, and CH₅₀ 49.3 U/ml. A test for HBs antigen was negative. Thyroid function tests were normal. Although urinary findings were normal, stools were positive for occult blood.

A roentgenogram of the chest showed no abnormalities. An MRI scan of the orbit revealed thickening of the right lateral rectal muscle, ipsilateral proptosis, swelling of the right optic nerve and enlargement of the retrobulbar space, though retrobulbar space-occupying lesions were not detected (Fig. 2). A CT scan of the orbit showed the proptosis, but no other abnormalities could be detected. A barium enema examination disclosed a sclerotic lesion on the descending colon that indicated ischemia or local inflammation. Celiac and renal angiograms showed small aneurysms in the liver and right kidney.

Two weeks after admission, the patient noted sensory loss corresponding to the distribution of the median nerve bilaterally. He lost 8 kg in the three weeks before treatment. Biopsy of the right gastrocnemius muscle was performed. The muscle biopsy specimen showed cellular infiltration involving a small artery and the surrounding connective tissue, with disruption of the elastic membrane.
He was diagnosed to have PN based on the criteria established by the Ministry of Health and Welfare, Japan (15). He was then immediately treated with methylprednisolone (1,000 mg, daily, i.v.) for three days, followed by 60 mg of prednisolone orally every day. After the pulse therapy, facial pain, abdominal pain, and muscle pain subsided, and facial swelling and exophthalmos improved gradually (Fig. 1, right). One month later, his sensory loss disappeared completely. Prednisolone was successfully tapered by 5 mg every two weeks to a maintenance dose of 10 mg/day. Now he is being followed in the outpatient service without any relapse.

Discussion

The present patient had many symptoms and signs that indicated the diagnosis of PN: pyrexia, weight loss, peripheral neuritis, myalgia, hematochezia, leukocytosis, radiologically multiple aneurysms, and histologically vasculitis with disruption of the elastic membrane. These symptoms completely fulfilled the diagnostic criteria for PN established by the Ministry of Health and Welfare, Japan, in 1990 (16), as well as those promulgated by the American College of Rheumatology in 1990 (17). The biopsy revealed transmural inflammation of a small artery without granulation or scaring, which indicated an acute inflammatory stage of the disease (16).

The present patient had an elevated value of GPT. This appeared to be a result of ischemic hepatitis secondary to intrahepatic arteritis, or direct damage due to arterial inflammation, because the aneurysms on the celiac angiogram indicated arteritis in the liver. The levels of the serum immunoglobulins prior to therapy were decreased; these normalized after therapy. Two reasons were suspected: consumption of immunoglobulins and a decrease in their production. But consumption was unlikely because the complement levels were within normal limits. Rather, the decreased production seemed to be due to abnormal immuno-regulation. The abnormalities of both GPT and immunoglobulins responded to therapy.

The patient presented with exophthalmos and facial swelling as the initial manifestations. These manifestations are uncommon in PN. Five cases with exophthalmos (5, 7–10), and only one case with facial swelling (11), secondary to PN have been reported. The combination of both found in this patient has not been previously reported. Furthermore, this is the first report in which an MRI scan was used for the study of exophthalmos secondary to PN.

The pathomechanism of exophthalmos and facial swelling in PN has not yet been fully elucidated. Bagegni et al reported a patient who presented with bilateral proptosis due to PN (5). The CT scan showed abnormal soft tissue behind both eyes. Orbital biopsy demonstrated acute vasculitis and fibrosis of the surrounding tissue. However, in other reports, the histologically-confirmed mechanical compression from the granulomatous tissue was suggested as the cause of exophthalmos in PN (9, 10). On the other hand, Ryan and Brady reported a...
case with unilateral facial swelling, and attributed the swelling to edema in the masseter muscles (11). In the present case, although the cause of neither exophthalmos nor facial swelling was histopathologically elucidated, the MRI scan disclosed thickening of the extraocular muscle and enlargement of the retrobulbar space. Both the visual impairment which was responsive to treatment and the swelling of the optic nerve on the MRI scan, may support the presence of intraorbital vasculitis. These findings suggest that intraorbital vasculitis similarly caused edema of the components in both the orbit and the surrounding subcutaneous tissue.

The MRI finding is not peculiar to the present case and is sometimes seen in other conditions, namely inflammatory orbital pseudotumor (18, 19). Therefore, it is impossible to reach a final diagnosis simply based upon the radiological studies. But MRI is helpful in confirming the mechanism of exophthalmos and in narrowing the differential diagnosis.

In summary, the present case indicates that systemic vasculitis, such as PN, can cause the unusual clinical manifestations of exophthalmos and facial swelling.

Acknowledgments: We are very grateful to Dr. J. Nakayama of the Department of Clinical Pathology, Shinsyu University, for his technical assistance and the histological analysis of the biopsy specimen.

References