A case of malignant atrophic papulosis with cranial nerve and peripheral nerve impairment*

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Abstract: Malignant atrophic papulosisis is a rare, multisystem obliterative vasculopathy of unknown etiology, occasionally involving the cranial nerve. We describe the first case of malignant atrophic papulosisis with cranial nerve and peripheral nerve involvement in China. A 47-year-old woman presented to our hospital with atrophic porcelain white papules over the trunk and extremities, numbness in the right calf, vision decrease and impaired movement of the right eye. She was diagnosed with malignant atrophic papulosisis, based on characteristic symptoms and histopathologic examination. The patient was treated with dipyridamole and aspirin for 9 months, but later died of gastrointestinal hemorrhage. We reviewed currently available case reports on cranial nerve involvement in malignant atrophic papulosisis and emphasized the importance of skin biopsy in diagnosing this disease.
Keywords: Cranial nerve injuries; Malignant atrophic papulosis; Peripheral nerve injuries

INTRODUCTION

Malignant atrophic papulosis (MAP) is a rare, multisystem obliterative vasculopathy of unknown etiology, occasionally involving the cranial nerve. We present a MAP case in a 47-year-old woman with cranial nerve and peripheral nerve involvement.

CASE REPORT

A 47-year-old woman presented to our clinic with a 6-year history of asymptomatic erythematous papules over the trunk and extremities. The lesions first appeared on her right leg and healed spontaneously, leaving an atrophic residual scar. Similar skin lesions had been appearing gradually and spreading across the whole body surface in the subsequent 5 years. At the age of 46, the patient experienced a 2-hour episode of visual field constriction in the right eye. Two months later, her right calf was anesthetic and she noticed a painless vision decrease and permanent visual field constriction in her right eye, which prompted her to seek advice.

Cutaneous examination revealed multiple, round and linear, atrophic porcelain white papules of 5-10mm in diameter over the trunk and extremities, with some showing an erythematous telangiectatic peripheral halo (Figure 1A). Neurologic examination found hypesthesia and paresis in the right calf. Cranial nerves examination showed that the abducent nerve and optic nerve were impaired, resulting in weakened movement on the right side, visual deterioration, and visual field constriction of the right eye (Figure 1B). Tendon reflexes were normal and Babinski signs were negative.

Histopathology revealed central epidermal atrophy, papillary dermal sclerosis, and perivascular lymphocytic inflammatory cell infiltrates, in addition to vascular thrombosis (Figure 2).

Analytical assessments were normal, including blood cell count, urinalysis, serum chemistry analysis, immunoglobulin profile, myocardial enzymes, CRP, ESR, RF and all plasma autoantibodies. But the fecal occult blood test was positive. Visual evoked potential

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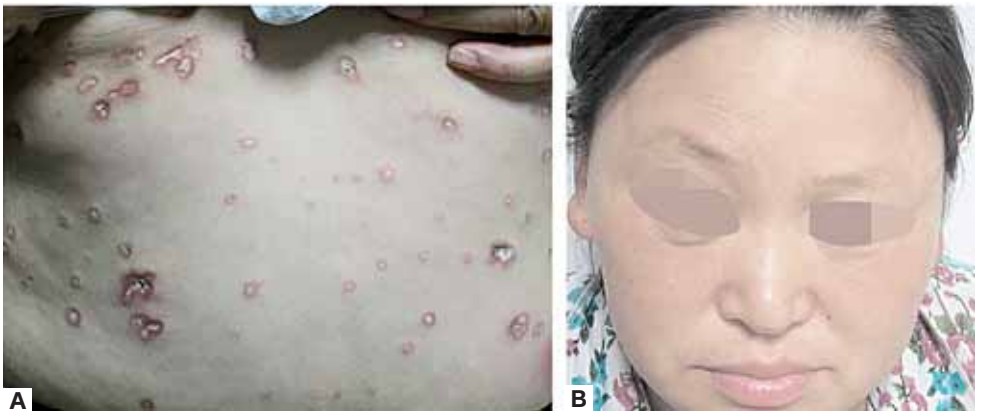

Figure 1: A. Cutaneous examination revealed multiple round and linear atrophic porcelain white papules of 5-10mm, with some showing an erythematous telangiectatic peripheral halo. **B.** Cranial nerves examination found impaired movement on the right side, visual deterioration, and visual field constriction of the right eye

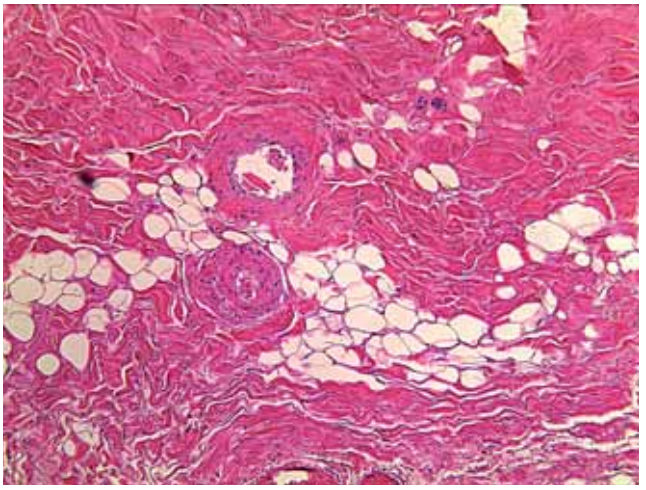

Figure 2: Skin biopsies revealed perivascular lymphohistiocytic infiltration. Vascular occlusion was also observed(HE×400)

showed the optic nerve was damaged. Ophthalmoscopic findings were normal. Gastroscopy examination uncovered chronic, gastric and duodenal inflammation. Screening colonoscopy demonstrated that the colon was congestive, puffy and erosive, with small, shallow ulcers. Chest computed tomography was normal. No intracerebral damage or venous thrombosis was found, as evidenced by brain magnetic resonance imaging and magnetic resonance angiography. Electromyologram (EMG) showed right quadriceps femoris neurogenic damage.

A diagnosis of MAP was made and the patient was treated with dipyridamole (50mg twice a day) and aspirin (100mg once a day) for 9 months, experiencing no symptoms, skin lesion improvement and no deterioration. The patient was lost to follow-up after 10 months. We later learned that she had died of gastrointestinal hemorrhage a year after presenting to our clinic.

DISCUSSION

MAP, also known as Degos disease, is a rare, multisystem obliterative vasculopathy of unknown etiology, which frequently affects the skin, gastrointestinal tract and central nervous system. Immunological dysfunction, hyper-coagulation, fibrinolytic disturbances, genetic predisposition and viral or streptococcal infection, may be involved in the pathogenesis of MAP.[1-3] Drawing on the dialogue 'Degos disease' between Noah Scheinfeld and Stuart Brown, Warren classified MAP into: classical MAP with systemic manifestations, further subclassified into autoimmune MAP, coagulopathy-associated MAP or virally-induced MAP; and benign cutaneous MAP.[4]

Our case is remarkable because of three unusual features. Firstly, physical examination revealed impaired function of cranial nerves, II and VI on the right side, which respectively manifested vision decrease and impaired movement on the right side of the right eye. Her skin lesions were neglected when she visited the department of neurology and ophthalmology. We reviewed currently available case reports on cranial nerve involvement in MAP; our patient was the world's fourth reported case (Chart 1).[5-7] In addition, some other ophthalmic signs have been reported in MAP, including conjunctival atrophic plaques, telangiectatic vessels, scleral thinning, blepharoptosis and optic atrophy. [6,8] Secondly, she repeatedly felt numbness in her right calf. EMG showed right quadriceps femoris neurogenic damage, which suggested the patient also had peripheral nerve involvement. Thirdly, although the patient had no clinical symptoms indicating gastrointestinal involvement, the fecal occult blood test was positive and colonoscopy showed that the colon was scattered with superficial ulceration, both of which prompted the diagnosis of intestinal involvement. In ancestral literatures, about 15% of MAP patients enjoy good health with their lesions limited to the skin.[9] Once multisystemic disease has developed, death occurs in approximately 50% of patients within

Chart 1: Patients' information statistics for cranial nerve involvement in MAP

Year reported	Source	Age	Gender	Period of disease progress before cranial nerve involved	Cranial nerve involved	Clinical Performances of cranial nerve involved	Therapy	Results	References
2009	Birmingham UK	6M	Male	3 weeks	Oculomotor nerve Abducent nerve	Disconjugate eye movements	Analgesics and antiepileptics	Died of multiple progressive cerebral infarctions	5
2006	Japan	58Y	Female	Initially	Optic nerve	Loss of vision in the left eye	Antiplatelet and anticoagulant	Died of septic shock	6
1992	Japan	41Y	Male	3 years	Facial nerve Abducent nerve	Facial hypesthesia	Heparin, urokinase, ticlopidine, dipyridamole, and prednisolone	Died of intracranial hemorrhages	7
2013	x country	47Y	Female	6 years	Optic nerve and abducent nerve	Impaired movement on the right side, visual deterioration, and visual field constriction of the right eye	Dipyridamole, aspirin	Died of gastrointestinal hemorrhage	Present case

one to two years. The most common causes of death are sepsis from peritonitis, CNS bleeding, and pleural or pericardial. [4]

There is no specific therapy for MAP. Some cases respond well to antiplatelets treatment. Darwich has reported a MAP case in which skin lesions improved significantly after treatment with chloroquine, aspirin and prednisone.[2] Magro has investigated four MAP cases and found prominent vascular C5b-9 in the skin, gastrointestinal tract and brain.[10] In all cases, they also found evidence of high interferon-α (IFN-α) expression. They proposed that C5 or IFN-α may emerge as potential candidate targets.❑

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