copy examination disclosed multiple nerve fiber layer infarcts in the posterior pole of the right eye with attenuated arterioles and segmentation of the blood column ("box-carring") and a contrasting red zone in the macula ("cherry-red spot"). The left fundus was normal. The erythrocyte sedimentation rate was 78 mm/1st hour.

Based on the clinical data, a diagnosis of GCA with arteritic central retinal artery occlusion was made. Therefore a right temporal artery biopsy was performed. Antiaggregation was maintained and treatment with methylprednisolone 1 g/i.v. over 2 hours followed by oral prednisone 60 mg/day was quickly started to avoid a possible bilateral visual loss. To our surprise, the following day, approximately 10 hours after the bolus of methylprednisolone, the patient's vision began to improve, initially in response to hand motions and then fairly rapidly thereafter. Five days after admission her vision was 20/30 in the right eye. At that time she was free of symptoms and the result of the right temporal artery biopsy was available and supported the diagnosis of GCA. During the follow-up her visual acuity has remained stable.

Permanent visual loss in GCA is generally due to anterior ischaemic optic neuritis (5, 6). Less commonly, visual loss may be secondary to central retinal artery occlusion, retrobulbar optic neuritis or even cortical blindness following an episode of vertebrobasilar stroke in the setting of GCA (6). Our experience with patients with visual loss was discussed in a collaborative and multicenter study of patients, more than 60% of whom resided in the Lugo area of northwestern Spain (6). In this regard, no improvement of visual loss was observed if the therapy was delayed by more than 2 days (6). However, in only 2 of our cases was permanent visual loss due to retinal stroke. Interestingly, visual recovery in patients with complete visual loss due to central retinal artery occlusion secondary to GCA has been previously reported (9, 10). However, the explanation for the visual recovery in our patient is unclear as the duration of arterial occlusion was longer than 48-72 hours. We may speculate that, besides the duration of visual loss, the degree of arterial occlusion may be another important factor for the recovery of vision. Indeed, it is possible that in our case profound but presumably incomplete retinal ischemia could explain the unexpected late improvement. In this regard, antiaggregation therapy might have been of some value in preventing complete arterial occlusion.

In summary, although it is difficult to find a satisfactory explanation for visual improvement after more than 3 days of retinal stroke, and there are no prospective double-blind controlled studies to counteract a putative prothrombotic effect of the vasculitis and supposedly of corticosteroids, we are in favour of considering antiaggregation therapy in such cases.

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A case of Bartter's syndrome, gout and Becker's muscular dystrophy

Sir,

Gouty arthritis associated with Bartter's syndrome is a very rare presentation. Generalized muscular weakness, muscle cramps or periodic paralysis often develop in patients with this disease because of chronic potassium depletion. This clinical picture may mimic conditions such as myopathy and muscular dystrophy. The association of Bartter's syndrome with Becker's muscular dystrophy has not been previously described. We present a patient with a highly unusual combination of Bartter's syndrome, Becker's muscular dystrophy and gout which regressed following appropriate therapy, and suggest that these medical conditions may be related.

A 49-year-old man with a 30-year history of muscular dystrophy was admitted to our hospital because of progressive shoulder and pelvic muscular weakness and painful swelling of the metacarpophalangeal and metatarsophalangeal joints of the left foot. The first signs of weakness had been difficulties in climbing stairs and recurrent falls, and the results of the diagnostic workup, which included myopathic electromyography (EMG) and muscle biopsy, were consistent with muscular dystrophy. The disease was slowly progressive.

More than 9 years before the current presentation, he was believed to have been affected by gout, based on acute attacks of arthritis and high uric acid levels confirmed by joint aspiration and crystal analysis (he had no risk factors for hyperuricemia). Past medical treatment consisted of allopurinol and colchicine. Before the current admission, he had experienced a significant progression of general weakness and had been frequently hospitalized for relapsing arthritis which worsened to the extent that he eventually became bound to a wheelchair. He denied taking diuretics or laxatives and there were no episodes of vomiting or diarrhea which could have caused the hypokalemia.

Physical examination revealed an afebrile man with blood pressure 120/80 mmHg and with greater weakness of the lower than the upper limbs. Deep tendon reflex responses ranged from decreased to absent. Other presenting features were pseudohypertrophy of the calf muscles, flexion contractures of the knees and elbows and severely deformed metacarpophalangeal and first metatarsophalangeal joints. Tophi were observed on the olecranon bursae, finger joints and helix of the ear.

Laboratory determinations showed: serum

potassium 2.7 mEq/L, Na 139 mEq/L, chloride 94 mEq/L, magnesium 2.03 mg/dl, uric acid 11.2 mg/dl, BUN 16 mg/dl, creatinine 0.7 mg/dl, creatine phosphokinase 777 mU/ml, and ESR 29 mm/hr. Blood gases were: pH 7.46, pCO_2 43.1 mmHg, bicarbonate 29 mEq/L. Complete blood count and protein electrophoresis were normal. Urinalysis showed pH 7.0, specific gravity 1.015, protein glucose negative, and no findings in the sediment. Serum potassium was 3.0 mEq/L and, after 3 days of sodium intake of approximately 120-150 mEq/day, the creatinine clearance was 86 ml/min. The 24-hour urine excretion of potassium was 118 mEq/day (FE 29%), sodium 113 mEq/day (FE 0.49%), chloride 103 mEq/day (FE 1.3%), magnesium 82 mg/day (FE 4.2%), uric acid 180 mg/day, and calcium 80 mg/day. Supine plasma renin activity was 23 ng/ml/hr (normal < 2), upright 25 ng/ml/hr (normal < 5), and supine plasma aldosterone 211 pg/ml (normal < 125). Thyroid function was normal. Tests for anti-DNA, SS-A, SS-B, antimitochondrial, antithyroglobulin, antimicrosomal and antiparietal cell antibodies were negative.

Direct molecular analysis of the dystrophin gene showed deletion of exons 45-47. Open biopsy of the left quadriceps muscle revealed groups of large, hypertrophic fibers around fibrotic areas and small atrophic muscle fibers including markedly reduced sarcoplasm and scattered angulated fibers with less extensive atrophic changes (Fig. 1). While immunostains to dystrophin showed dystrophin localized to the sarcolemma of the all muscle fibers, the patient's fibers were less intensely stained than normal. In addition, there were minor staining gaps around some of the fibers.

The EMG showed myopathic changes with early recruitment. Indomethacin and potassium supplementation were given and led to a gradual improvement of musculoskeletal symptoms, while serum potassium increased to a maximum value of 3.5 mEq/L.

Bartter's syndrome does not represent a unique entity but encompasses a variety of disorders of renal electrolyte transport. In the present case report, both the clinical features and the laboratory determinations are consistent with the diagnosis of Bartter's syndrome. The molecular defects were defined as renal bumetanide Na-K-2Cl (NKCC2) cotransporter and ATP-sensitive inwardly rectifying K channel (ROMK), and a deletion or mutation at gene encoding a renal chloride channel (ClCKb) was identified (1-4). Muscle biopsy, DNA studies, EMG and the creatine phosphokinase confirmed the diagnosis of Becker's muscular dystrophy.

Our patient had symptomatic hyperuricemia. Gout is the most common cause of inflammatory arthritis in males over 40 years of age. Hyperuricemia may be associated with many medical conditions including Bartter's syndrome (6, 7). Meyer *et al*. reported the presence of hyperuricemia in 50% and gouty arthritis in 20% of a group of patients with BS (5). Systemic alkalosis was considered as a cause of decreased uric acid clearance, as had been found in our patient. However, the contribution of plasma volume contraction as part of the syndrome in the case of hyperuricemia cannot be excluded.

Increased PGE2 synthesis resulting from hypokalemia has been reported in Bartter's syndrome (8). In some cases, indomethacin does reduce potassium wasting but in our case (together with potassium supplementation), it had a limited effect.

We present an unusual case of coexistent gout, Bartter's syndrome and Becker's muscular dystrophy. The nature of this relationship is unknown and their occurrence in a single patient remains unexplainable, and may be no more than a coincidence. Muscular weakness is a manifestation of both Becker's muscular dystrophy and Bartter's syndrome. While no specific therapy is available for the slowly progressive weakness of muscular dystrophy, some beneficial effect can be obtained by potassium supplementation and prostaglandin synthesis inhibitors.

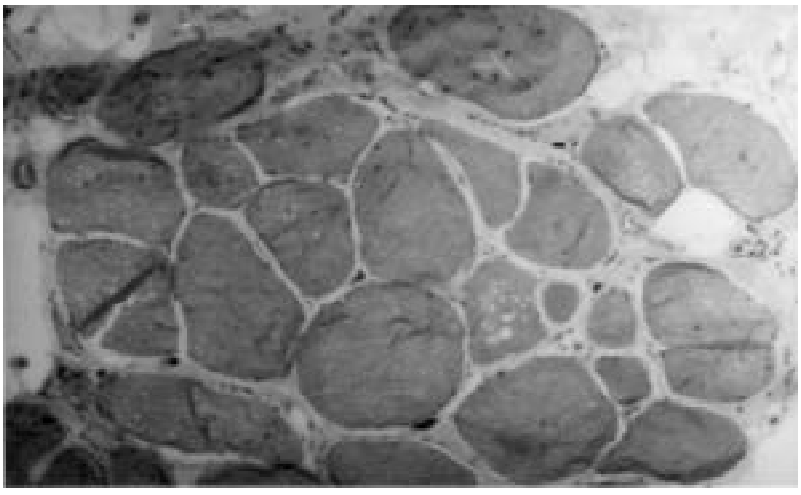

Fig. 2. A left quadriceps muscle biopsy showing groups of large, hypertrophic fibers around the fibrotic areas and small atrophic muscle fibers.

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