Immunoglobulin A vasculitis (IgA vasculitis [IgAV]; formerly known as Henoch Schönlein purpura [HSP]) is one of the most common systemic vasculitic diseases in childhood. IgAV is an immune-mediated leukocytoclastic small vessel vasculitis characterized by non-thrombocytopenic, palpable purpura on the lower extremities. It is a multisystem disease that typically affects the skin, joints, gastrointestinal tract and kidneys.¹ Nervous system involvement is uncommon. Headaches, seizures, visual changes and reduced conscious levels are the most frequent neurologic symptoms. Peripheral nervous system dysfunction has been reported more rarely.² Here we report an 11-year-old boy who was diagnosed as IgAV and developed axonal degeneration of the left tibial nerve during follow-up.

Case Report

An 11-year-old boy was admitted to our clinic with pain and swelling in both his ankles as well as rashes. His past and family history...
were not remarkable. His blood pressure was normal. Physical examination revealed edema and tenderness in both ankles and symmetric palpable purpura extending from the knees to the dorsum of his feet. Laboratory findings were as follows: hemoglobin 13 g/dl, white blood cell count 13,000/mm³, platelet count 251,000/mm³, erythrocyte sedimentation rate 13 mm/hour, C-reactive protein 12.5 mg/L, and ASO 467 IU/ml. A throat culture was negative for group β-hemolytic Streptococcus. Urinalysis, blood biochemistry, serum immunoglobulin levels, C3 and C4 levels were normal. An investigation of hepatitis and viral markers, antinuclear antibody, anti-double stranded DNA and antineutrophil cytoplasmic antibody (ANCA) were all negative.

IgAV diagnosis was established according to European League Against Rheumatism (EULAR)/Paediatric Rheumatology International Trials Organisation (PRINTO)/Paediatric Rheumatology European Society (PRES) criteria with palpable purpura and arthritis. A skin biopsy was performed; it showed leukocytoclastic vasculitis, with IgA deposition. Bed rest and non-steroidal anti-inflammatory treatment was recommended and out patient follow-up was started. On the 18th day of follow-up, he was admitted with widespread palpable purpura, myalgia and edema in the lower extremity, abdominal pain and left scrotal swelling. The patient was hospitalized and intravenous prednisolone 2 mg/kg/day was started due to scrotal and abdominal involvement. All symptoms improved and edema was resolved, but on the 3rd day of the prednisolone therapy, the patient suffered from numbness in the left foot. Neurological examination revealed hypoesthesia in the soles of the left foot and the thumb. Deep tendon reflexes were normal and he had no motor abnormalities. Electromyoneurography (EMNG) was performed; it showed moderate to severe axonal degeneration of the left tibial nerve. Bolus methylprednisolone (30 mg/kg/day) was given for 3 consecutive days, followed by oral prednisolone, but the hypoesthesia in his left foot continued. Although intravenous immunoglobulin (IVIG) 1 g/kg/day for 2 consecutive days was given, the patient’s symptoms didn’t improve. Cyclophosphamide bolus was given (500 mg/m²) seven days after IVIG treatment. All of the patient’s neurological complaints and signs regressed within one week and he was discharged. His oral prednisolone was gradually tapered and stopped at the end of the third month. After a follow-up period of 1 year, the patient had no complaints. Control EMNG was not performed due to improvement of clinical findings after treatment. The patient had no history to suggest familial Mediterranean fever (FMF) and MEFV gene analysis was found to be normal.

A written informed consent was obtained from the patient.

Discussion

Immunoglobulin A vasculitis is the most common systemic vasculitis of childhood. The diagnosis of IgAV is based on clinical criteria and histopathological findings. At the first admission, our patient was diagnosed as IgAV with typical palpable purpura and arthritis according to the EULAR/PRINTO/PRES classification criteria which were validated specifically for childhood-onset disease. Approximately 3 weeks later, while he was hospitalized on steroid treatment for gastrointestinal and scrotal involvement, peripheral neuropathy developed which is a finding that is also very rarely reported in IgAV. Other systemic vasculitis should be considered in differential diagnosis of a patient with purpura and neuropathy. Skin biopsy findings are also important to exclude other forms of vasculitis such as ANCA-associated vasculitis. Our patient’s skin biopsy revealed in leukocytoclastic vasculitis, with IgA deposition. Moreover, negative ANCA tests, absence of renal involvement, no relapse during follow-up after discontinuation of steroid treatment all ruled out especially the diagnosis of microscopic polyangiitis.
Belman et al. reported that nervous system involvement in IgAV is usually underestimated. Garzoni et al. reviewed nervous system dysfunction in IgAV and showed that peroneal neuropathy, peripheral facial palsy, Guillain–Barre’ syndrome, brachial plexopathy, posterior tibial nerve neuropathy, femoral neuropathy, ulnar neuropathy and mononeuropathy multiplex were reported as cranial or peripheral neuropathy conditions. It has been suggested that deposition of circulating immune complexes containing IgA on myelin sheaths could cause the peripheral demyelination in IgAV. Furthermore, systemic vasculitis may cause infiltration of the vasa nervorum or the epineurial arteries by inflammatory cells resulting in ischemia and thrombosis of the peripheral nerves. However, sometimes mechanical compression of the nerve because of local edema and joint effusion may cause peripheral neuropathy. Although our patient’s edema was resolved with steroid treatment, his neurological complaints continued. Inflammatory infiltration of small vessels and ischemia probably resulted in axonal degeneration of the left tibial nerve of our patient.

There is no a clear treatment recommendation for peripheral neurological involvement in IgAV. Patients were reported to improve following bolus methylprednisolone administration and IVIG treatment should be given in IgAV complicated by acute inflammatory neuropathy like Guillain-Barre syndrome. Our patient’s clinical course didn’t get better after bolus corticosteroid treatment and IVIG was administered. Therefore, for the possibility of a permanent sequelae in our patient we gave a single dose of bolus cyclophosphamide treatment and the symptoms began to improve and disappeared within one week.

Although peripheral neuropathy is a rare complication of IgAV, it may have a resistant course. Cyclophosphamide therapy should be kept in mind in patients who don’t respond to intensive steroid therapy and IVIG.

**Author contribution**

The authors confirm contribution to the paper as follows: study conception and design: FA, BÇA; data collection: TK; analysis and interpretation of results: ZET, EÇ; draft manuscript preparation: FA, BÇA. All authors reviewed the results and approved the final version of the manuscript.

**Conflict of interest**

The authors declare that there is no conflict of interest.

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