Spontaneous spinal subdural hematoma as a manifestation of ANCA Associated Vasculitis: description of three cases

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Objectives

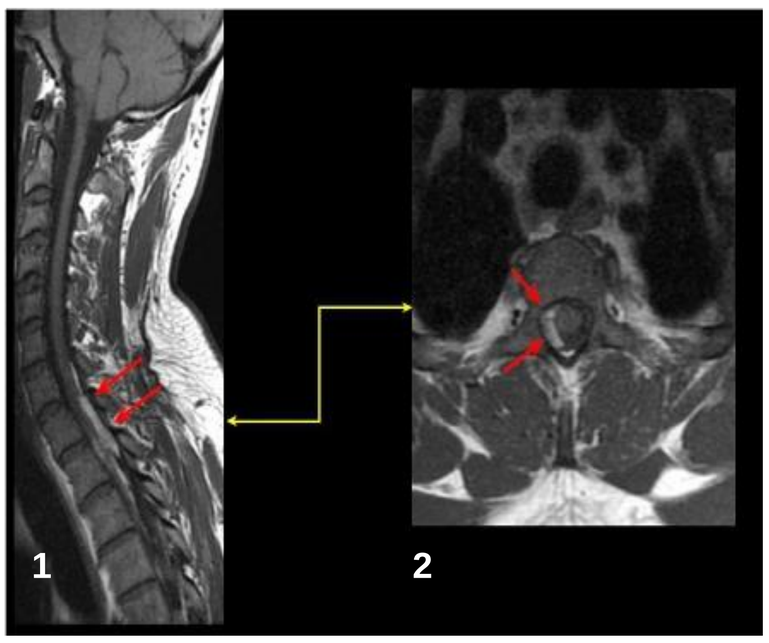
Spinal hematoma (SSH) is a rare but serious condition, prevalently seen in neurological emergency departments. We report three cases of spontaneous SSH in patients with antineutrophil cytoplasmic antibody Associated Vasculitis (AAV).

Materials and methods

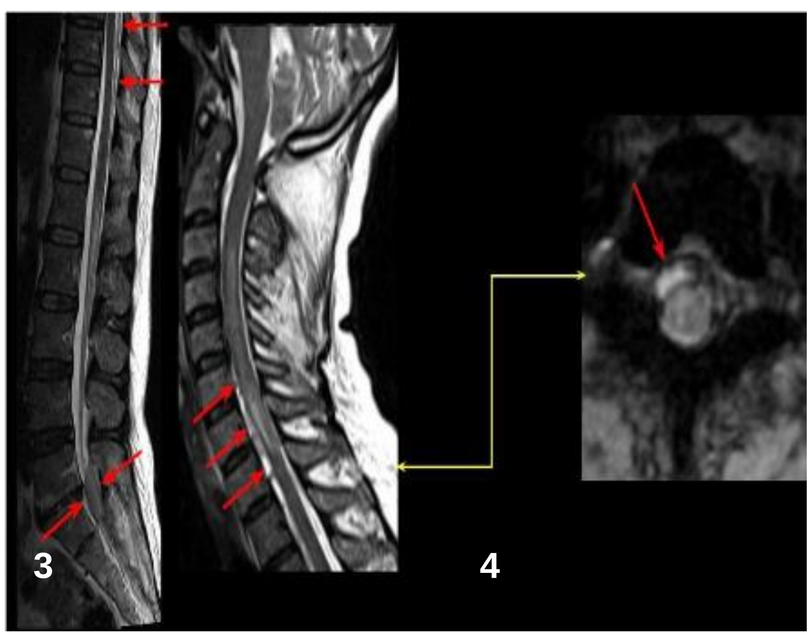
Case 1 a 40 years- old man with a history of vasomotor rhinitis and asthma, was in –hospitalized due to sudden appearance of fever, arthromyalgias and cervical-dorsal back pain. Blood exams showed hypereosinophilia, high values of c-reactive protein, creatine phosphokinase and Anti Myeloperoxidase Neutrophil Cytoplasmic Antibody (MPO). Spine magnetic resonance imaging showed a cervico-dorsal spinal subdural hematoma. Symptoms improved with intravenous steroid therapy (intravenous Methylprednisolone) for 5 days.

Case 2 a 55 year old woman was in-hospitalized due to nausea, vomiting and painful left cervico-brachial syndrome. Laboratory test revealed chronic kidney disease stage V. Blood exams showed anti-MPO ANCA positivity. Extracorporeal dialysis with heparin was promptly started. The day after he developed acute flaccid tetraplegia with neurological bladder. Spine MRI showed cervico-thoracic subdural hematoma with secondary myelopathy for which she underwent surgical decompression. Although the patient was treated with plasmapheresis, Rituximab and intravenous steroid, her renal function didn't improve.

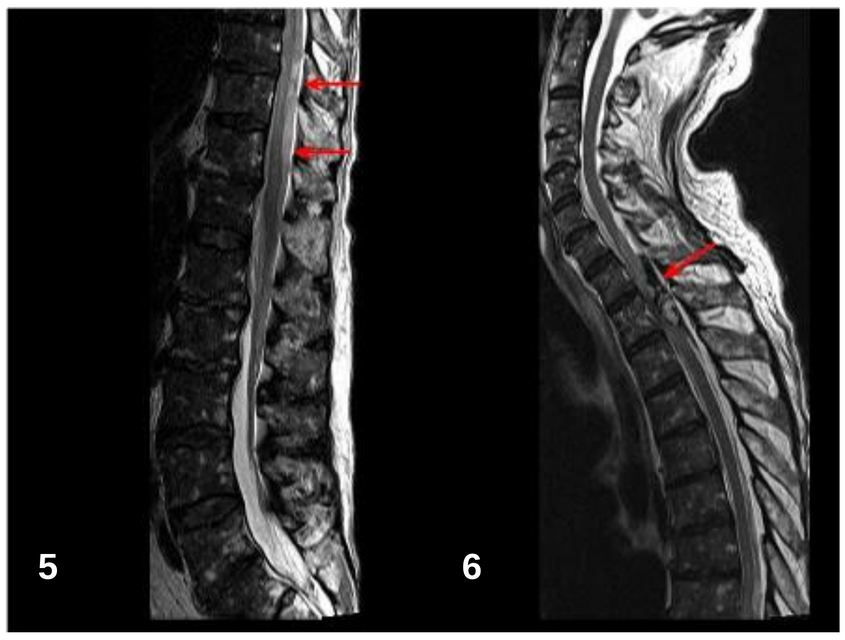
Case 3 a 67 year old man with a four months history of fever, arthralgias, weight loss, left pleural effusion and high titre Anti-proteinase 3 antibodies, monoclonal gammopathy, was in-hospitalized due to lobar pneumonia. He had a history of professional exposure to mineral silicates. Deep venous thrombosis of the left lower limb was also diagnosed and anticoagulation with heparin was promptly started. After 36 hours he developed a flaccid paraplegia and bilateral upper arms paresis. Spine MRI confirmed the presence of SSH at C6-T1, for which he underwent surgical decompression. Brain computerized tomography showed bilateral posterior stroke, complicated by structural epilepsy. Vasculitic manifestations went into complete remission thanks to intravenous Desamethasone and Rituximab.



Figures 1-2: case 1 sagittal and axial spine views



Figures 3-4: case 2 sagittal and axial spine views



Figures 5-6: case 3 sagittal spine views

RESULTS: we performed in all patients diagnosis of AAV. Only one of them performed complete neurological recovery (Case 1) while neurological examination of cases 2-3 showed residual upper arm paresis and lower arm paraplegia.

Conclusion

Diagnosis of AAV should be considered in presence of spontaneous spinal subdural hematomas with neurological quickly worsening signs. Concomitant anticoagulation therapy may worsen course and prognosis, so we should perform cost-benefit analysis in administering anticoagulants and in establishing the dose. Professional exposure to mineral silicates can have a role in the etiology of AAV so it should be taken into consideration.

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