척수재활

게시일시 및 장소 : 10 월 18 일(금) 13:15-18:00 Room G(3F)

질의응답 일시 및 장소 : 10 월 18 일(금) 16:17-16:21 Room G(3F)

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IgG4-Related Sclerosing Pachymeningtis: Report of Three Cases

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Immunoglobulin G (IgG) 4- related sclerosing disease is recently understood as an immune mediated fibroinflammatory condition that can involve various organs. Any other organ can be involved but the most common organ is Pancreas.IgG4 Related sclerosing disease is known for a systemic disease characterized by mass forming inflammatory lesions with abundant infiltration of IgG4-positive plasma cells and T-lymphocyte infiltration. There have been a number of reports of extrapancreatic cases. However, CNS involvement cases have been unusual. Among those rare cases, the most commonly reported CNS lesion is the pituitary gland accompanied with hypopituitarism, diabetes insipidus and/or local mass effect. Pachymeningitis is a chronic progressive diffuse inflammatory fibrosis of the dura mater. We report three cases of IgG4-related sclerosing pachymeningitis which involves spinal cord causing weakness of limbs and pain. There is no international standard for the histological diagnosis of extrapancreatic IgG4-related disease. Biopsy can be used to rule out other condition first; then serum IgG4 level could be helpful and corticosteroid therapy can be initiated to confirm the diagnosis. Histopathological finding of biopsy specimens is essential in the diagnosis of IgG4-related disease. IgG/IgG ratio > 0.4 and >10 IgG4 positive cells per High-Power Field (HPF) are supportive of IgG4-related disease, but neither one is a specific diagnostic marker. Correlation with specific histopathological finding is crucial regardless of serum IgG4 concentration, the number of IgG-positive plasma cells, or the ratio of IgG4 to IgG in tissue. The major histologic features of IgG4-related disease are a dense lymphoplasmacytic infiltration characterized a storiform (i.e., irregularly whorled) fibrosis, obliterative phlebitis and an eosinophil infiltrate. IgG4 related sclerosing disease responds well to steroid therapy. The disease is often mistaken for malignancy (ex. lymphoma), an infection or other immune mediated disease such as Sjogren's syndrome. Also, the disease could occur in multiple organs concurrently. Therefore, it is important to recognize IgG4-related conditions so patients do not go through unnecessary surgery or chemotherapy and could be treated with Steroid as soon as possible. We report rare cases of IgG4-related sclerosing disease involving spinal dura mater and found other organs involved as well in two cases. To find other sites of involvement, PET CT could be performed like our cases. There were some differences in out cases from former reported cases. Case patients did not respond well to steroid therapy. Patients are still taking Prednisolone or other types of immune suppressive medication. All of case patients have some sequelae e.g. pain, paresthesia and in severe

cases, weakness is still existed. Also other organ involvement was shown simultaneously or with some time gap. IgG4-related sclerosing disease should be see other organ involvement.

Table 1. Study cases: clinical presentation and pathologic features

	Case1	Case2	Case3
Sex/age	F/54	M/43	F/60
Clinical presentation	Right buttock, posterolateral thigh pain	Both arm and leg weakness	Posterior Neck pain, Lt shoulder pain, bilateral U/Ex radiating pain, Below T10 Hypesthesia, urinary symptom
Site of lesion	L5-S1	C4-6	C3-T4
Serum IgG4(mg/dL)	18.9	10.8	27.2
Serum IgG(mg/dL)	1286	1314	672
other organ involvement	none	Tonsil	Gallbladder
respond to steroid therapy	Good	Poor	Poor
pathologic findings	Chronic inflammation with lymphoplasmacytic infiltration and fibrosis	Fibrous tissue with lymphocytic infiltration	Chronic inflammation with lymphoplasmacytic infiltration and fibrosis
IgG4+ Plasma cells(/HPF)	up to 40	Positive in a few plasma cells	up to 7
IgG4/IgG Ratio	0.2	0.1	0.25

IgG; Immunoglobulin G, IgG4; Immunoglobulin G4; HPF; high-power field

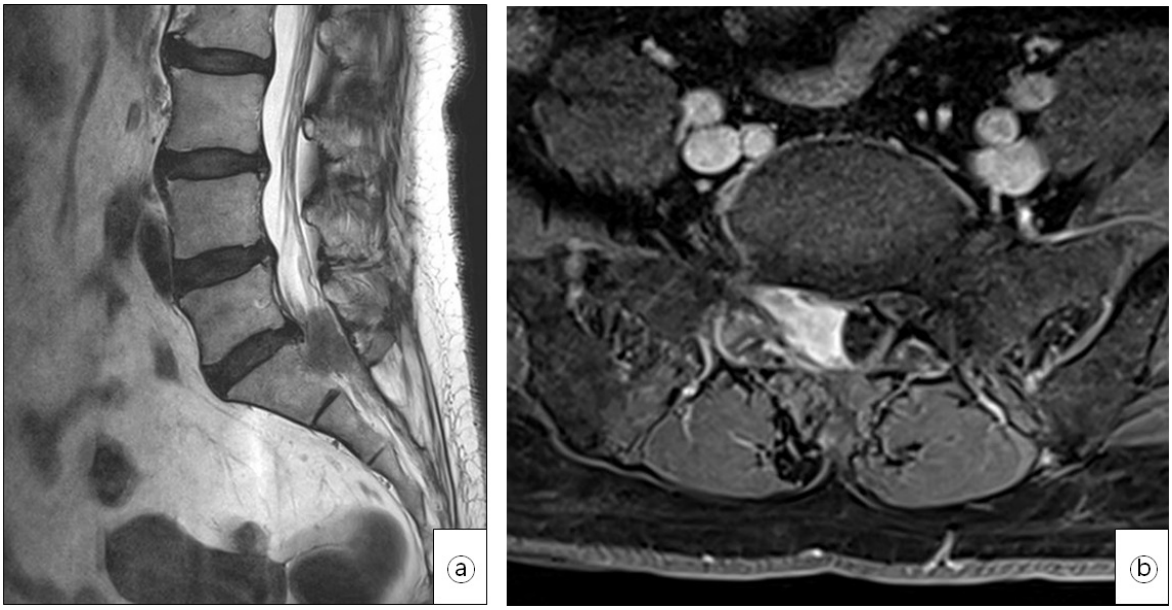

Figure 1. Sagittal T2-weighted image show a hypo-intense mass in the spinal canal that extends from L5 to S1.T1-weighted image with fat suppression reveals attenuated enhancement of the mass, which is predominantly located in right epidural space so epidural sac pushed to left.

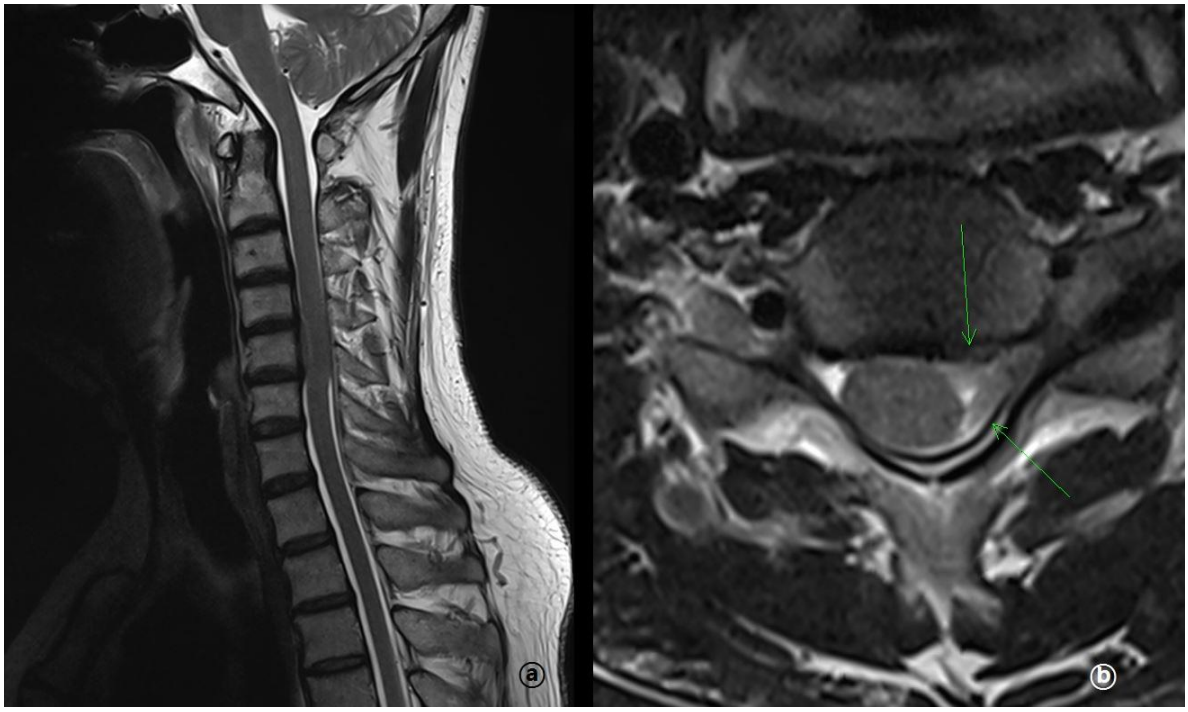

Figure 2. Sagittal T2-weighted (A) and axial enhanced T1-weighted (B) MRI, showing extent of pachymeningitis involving left side of epidural space from C4 to C6 level.