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Hypertrophic pachymeningitis IgG4 related, a rare presentation

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Abstract : Extraaxial pachymeningeal enhancement may arise from various benign or malignant processes, including transient postoperative changes, intracranial hypotension, neoplasms such as meningiomas, metastatic disease (from breast and prostate cancer), secondary CNS lymphoma, granulomatous disease or idiopathic.Hypertrophic pachymeningitis (HP) is an inflammatory condition in which the dura mater of the cranium or spine becomes thickened, leading to symptoms that result from mass effect, nerve compression, or vascular compromise.. Many times, no diagnosis is reached in such cases, the disease has been described as idiopathic HP. IgG4-related disease (IgG4-RD) is a recently described inflammatory condition known to cause tumefactive lesions at myriad anatomical locations.Here we present a case of IgG4 related pachymeningitis-a rare presentation

Keyword : IgG4 related diseases, IgG4 related hypertrophic pachymeningitis

INTRODUCTION

Extraaxial pachymeningeal enhancement may arise from various benign or malignant processes. Here we are presenting a case with interesting etiology with pachymeningeal enhancement.The cases were investigated by using a Siemens 3T Magnetom Skyra

History

41 year old female, presented with progressive compressive myelopathy and paraplegia for one month, the patient had no history of trauma or fever On clinical examination,the power was 3/5 in both lower limbs.Cerebrospinal fluid (CSF) analysis after lumbar puncture showed normal cell counts and glucose levels.Microbiological evaluation of the CSF for VDRL, and cultures for acid fast bacilli (AFB) and was negative. X ray and CT of dorsolumbar spine detected no significant abnormality. So the patient was referred for MRI of dorsolumbar spine

MRI findings:

Long segment extraaxial T2 hypointense lesion involving the dura in the dorsal segments of the cord(as in fig1).The lesion was also T1 hypointense(as in fig2)and showed no diffusion restriction. The spinal cord was normal and showed no abnormal signal intensities.On gadolinium scans,the extraaxial lesion showed diffuse homogenous enhancement of the thickened duramater(as in fig3). No abnormal enhancing lesions within the spinalcord were noted.Axial T2W images show non stenotic aortic wall thickening(as in fig4)

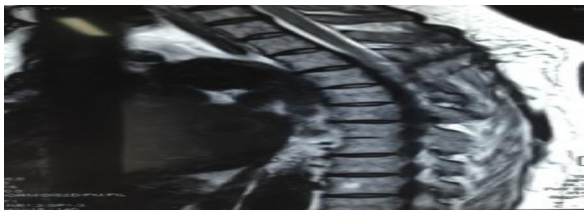

Fig1-Extraaxial hypointense lesion involving the duramater in the dorsal segments(T2W)

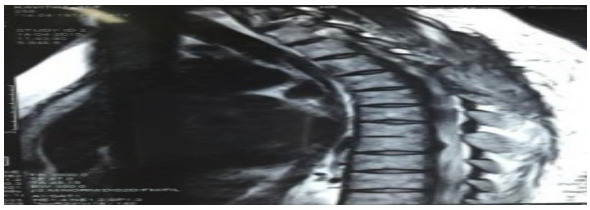

Fig2-Extraaxial hypointense lesion involving the duramater in the dorsal segments(T1W)

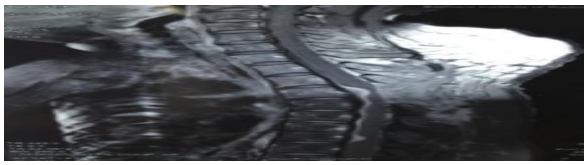

Fig3-Extraaxial hypointense lesion involving the duramater in the dorsal segments showing diffuse homogenous enhancement

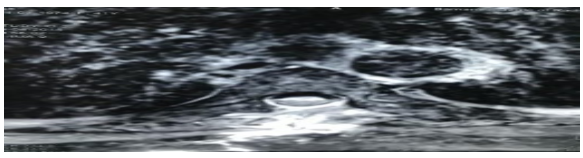

Fig4-Axial T2W-Aortic wall thickening

Differential diagnosis of pachymeningitis including sarcoidosis, tuberculosis meningitis, rheumatoid arthritis, syphilis, and occult malignancy, was sought and was excluded with negative laboratory results and absence of related clinical symptoms. Routine urine analysis, blood counts and serum chemistry were normal, except for an elevated erythrocyte sedimentation rate (ESR) of 55 mm/hr. The serum was negative for rheumatoid factor, antinuclear antibodies, VDRL test, hepatitis B surface antigen (HBsAg) and anti-double stranded DNA. Work up for sarcoidosis such as chest radiograph, serum calcium were negative. ultrasound scan of the abdomen, to exclude a neoplastic and chronic inflammatory processes, were normal. Cerebrospinal fluid (CSF) analysis after lumbar puncture revealed 2 lymphocytes/mm,3 protein of 26 mg/dl and sugar of 78 mg/dl. Microbiological evaluation of the CSF for VDRL, and cultures for acid fast bacilli (AFB) and fungi yielded no positive results The patient was diagnosed to be a case of idiopathic hypertrophic pachymeningitis. However literature revealed not all cases of idiopathic hypertrophic pachymeningitis were idiopathic, some were related to a spectrum IgG4 related disorders. Serum IgG4 levels for this patient was done and was found to be elevated(180mg/dl).N<135mg/dl.The patient also had non stenotic irregular aortic wall thickening, a finding in IgG4 related sclerosing disease along the aorta The patient was diagnosed to be a case of IgG4 related hypertrophic pacymeningitis. The patient was started on steroids and showed marked clinical improvement.

Discussion:

Pachymeningitis is defined as localised or diffuse thickening of duramater, leptomeninges, tentorium.

The causes include Infection- TB , fungal,Autoimmune diseases-wegner granulomatosis, rheumatoid arthritis, Intracranial hypotension,Malignancy – dural carcinomatosis, metastatic deposits, meningioma,Post surgical-VP shunt,Idiopathic hypertrophic pachymeningitis. Some of the cases descibed as idiopathic,belonged to IgG4 related pachymeningitis IgG4 related diseases are an important cause of diffuse pachymeningeal thickening and must be considered a important differential diagnosis .The 2 main criteria are 1) serum IgG4 concentration >135 mg/dL, and 2) >40% of IgG+ plasma cells being IgG4+ and >10 cells/high-powered field of biopsy sample in any affected organ. Manifestations of IgG4 related autoimmune diseases include enlargement of salaivary glands and lacrimal glands(mickulicz disease), pituitary lesions (hypophysitis), thickening of dura(pachymeningitis), thyroid lesions(Hashimotos thyroiditis, reidels thyroiditis ,autoimmune pancreatitis, sclerosing cholangitis, orbit(orbital pseudutumor), vessel wall thickening.In MR imaging, marked T2 shortening(T2 hypointensity), presuambly related to fibrosis and sclerosis In the head and neck and brain, manifestations of IgG4-RD include enlargement of salivary glands and lacrimal glands.CNS involvement in IgG4-related sclerosing disease includes infundibulohypophysitis, hypertrophic pachymeningitis, pansinusitis, inflammatory orbital pseudotumor, and chronic subdural hemorrhage.pathologic studies of pituitary lesions associated with IgG4-RD have reported fibrosclerosis with infiltration of lymphocytes and plasma cells. infundibulohypophysitis" of the anterior and posterior lobes of the pituitary gland was observed, leading to enlargement of the pituitary stalk and gland. Thus, lymphocytic hypophysitis may be due to IgG4-RD in some cases, and appropriate laboratory tests for IgG4 should be carried out for this condition IgG4-Related Autoimmune Pancreatitis-pancreas is either diffusely or focally enlarged with irregular narrowing of the main pancreatic duct and, often with stenosis of the common bile duct. Over time, atrophy occurs. loss of normal pancreatic clefts and a peripheral smooth (capsulelike) rim of attenuation have been described as characteristic signs .

Imaging findings in combination with increasedlevels of IgG and a conjoint appearance withother autoimmune diseases should raise suspicion of IgG4-related autoimmune pancreatitis. Signal intensity on T1 and T2 weighted MR images is nonspecific *Hepatobiliary Tract Involvement*- Both the intrahepatic and extrahepatic segments can be involved with focal or diffuse bile ductal wall thickening, mostly associated with stenosis and upstream cholestasis. Tumorlike infiltration involving the cystic, hepatic, choledochal, and small intrahepatic bile ducts. initially this may suggest cholangiocarcinoma .In cholangiocarcinoma, tumor infiltration is usually confined to either extrahepatic or intrahepatic bile ducts, frequently leading to focal cholestasis and liver capsule retraction due to desmoplastic growth. Liver involvement is more common in the form of a mass lesion at the hepatic hilum that encases the bile ducts but may also be manifested by diffuse infiltration of the periportal spaces (IgG4-related hepatopathy) *Lacrimal and Salivary Gland Involvement*-The lacrimal and salivary glands are commonly involved in IgG4-related sclerosingcdisease, presenting with unilateral or bilateral diffuse or focal swelling. At imaging,the lacrimal gland is enlarged and homogeneously enhancing, but these findings are nonspecific In the parotid and submandibularglands, sialadenitis can be focal and nodular in character, mimicking neoplasia. Isolated involvement of the submandibular gland (Kuttner tumor) presents as a firm swelling difficult to differentiate from a neoplasm *Retroperitoneal Fibrosis*-Retroperitoneal fibrosis is a manifestation of IgG4-related sclerosing disease resembling other types of fibrosis or vasculitis. Unlike giant cell vasculitis, IgG4-related sclerosing disease is usually located along the abdominal aorta and iliac vessels, presenting as nonstenotic vessel wall thickening with irregular margins In early disease stages, marked contrast enhancement

Later, however, fibrosis predominates *Renal Involvement* -Renal involvement is common in IgG4-related sclerosing disease and predominantly involves the cortex . Renal parenchymal lesions usually appear as bilateral peripheral cortical nodules, well-defined wedge-shaped lesions, or diffuse involvement. Images may show cortical nodules coalescing to a perirenal rim of enhancing tissue . Enlargement of both kidneys is also frequent and reflects tubulointerstitial nephritis. In the more focal infiltration pattern, multiple bandlike or broad-based triangular lowsignal-intensity areas are seen in the peripheral cortex of both kidneys. The main differential diagnoses are lymphoma, which generally is not seen in a symmetric, perirenal distribution *Lymph Node Involvement*-Lymph node involvement is detected on imaging studies in 80% of cases of IgG4-related sclerosing disease and may even be the initial manifestation . It is believed that lymph node enlargement does not represent true IgG4-related sclerosing disease but rather reflects the immune disturbance accompanying this disease . Focal or disseminated node enlargement with diffuse marked enhancement is usually found at imaging *Pulmonary Involvement* -Pulmonary involvement in IgG4-related sclerosing disease consists of multiple lung nodules, peribronchiolar infiltration , round ground -glassopacities, or reticulointerstitial or hilar adenopathy .Differentiation from pneumonia, sarcoidosis , amyloidosis may be difficult and usually requires biopsy

Conclusion

IgG4 related diseases may involve multiple organs with varying clinical presentations depending on the organ of involvement. Here we have presented a case which presented with paraparesis and had diffuse pachymeningeal thickening . In a patient presenting with pachymeningeal thickening which is T2 hypointense and showing contrast enhancement, a possible diagnosis of IgG4 related should be considered IgG4 related disease shows dramatic response to steroids and familiarity with clinical and imaging manifestations avoids a delay in diagnosis and unnecessary invasive interventions Cognizance of the broad spectrum of IgG4-related sclerosing disease should raise the level of suspicion and prompt a search for systemic manifestations of this disorder and thus avoid misdiagnosis with consecutive unnecessary surgical procedures .

References

Idiopathic Tumefactive Hypertrophic Pachymeningitis Imran A. Kazem, MD, Natasha L. Robinette, MD, Norbert Roosen, MD, Michael F. Schaldenbrand, MD, and Joon K. Kim, MD

MR Imaging of IgG4-Related Disease in the Head and Neck and Brain K . Toyodaa, H. Obaa, K. Kutomi a , S. Furuia, A. Ooharab, H. Moric, K. Sakuraid,K. Tsuchiyab, S. Kane and Y. Numaguchif

Systemic IgG4-Related Sclerosing Disease: Spectrum of Imaging Findings and Differential Diagnosis-Marius Horger1 Hans-Georg Lamprecht2 Roland Bares3 Daniel Spira Marc Schmalzing4 Claus Detlef Claussen1 Patrick Adam5(AJR)

IgG4-related disease and hypertrophic pachymeningitis.- Wallace ZS[1], Carruthers MN, Khosroshahi A, Carruthers R, Shinagare S, Stemmer-Rachamimov A, Deshpande V, Stone JH

• IgG4-related Sclerosing Disease: Autoimmune Pancreatitis and Extrapancreatic Manifestations Paraskevi A. Vlachou, MD , Korosh Khalili, MD , Hyun-Jung Jang, MD Sandra Fischer, MD ,Gideon M. Hirschfield, MD , Tae Kyoung Kim, MD.

IgG4-related Disease from Head to Toe-Anxo Martínez- de-Alegría, MD, SandraBaleato-González, PhD, Roberto García-Figueiras, PhD, Anaberta Bermúdez-Naveira, MD,Ihab Abdulkader-Nallib, MD, José A. Díaz-Peromingo, MD, Carmen Villalba-Martín, MD .

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