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10-27-2020

Idiopathic Hypertrophic Pachymeningitis in the Setting of Chronic Cutaneous Lupus Erythmatosus: A Case Report

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Introduction

Idiopathic hypertrophic pachymeningitis (IHP) is a rare disease when no other vascular abnormality, infection, trauma, neoplasm and neuro-inflammatory condition exists. It causes chronic inflammation with fibrosis of the cerebral and/or spinal dura mater [2].

Clinical features include headache, cranial nerve impairment, paraparesis, ataxia, hydrocephalus, papilledema, hemorrhage, or secondary vascular occlusions [3]. Magnetic resonance imaging (MRI) is essential for diagnosis. Most common sites of localization are cranial (79%), spinal cord (15%), and craniospinal (5.5%) [4,1].

Case Report

51-year-old Hispanic female with diagnosis with SLE twenty years prior based on a clinical presentation with a malar photosensitive rash, oral sores and Raynaud's disease.

In 2015 Brain MRI revealed a 1.5 x 1.5 x 0.9 cm enhancing posterior fossa mass noted as an incidental finding and possible meningioma, however patient did not follow up on discharge.

In Early 2019, she presented to rheumatology with complaints of sharp pressure-like neck pain, worse with lying down and extension, neck stiffness, difficulty initiating movement of the neck, associated with numbness in her posterior neck and left upper extremity as well as balance difficulties. MRI cervical spine revealed an extra-axial dural mass measuring 5.8 cm in length by 1 cm anteroposterior by 2.5 cm transverse; progressively enlarged when compared to 2015 (see Figure 1A and 2A). She had neurosurgical intervention and biopsy on final pathologic report was positive for hypertrophic pachymeningitis. The mass showed microscopic sections of myelofibrosis, predominantly perivascular lymphohistiocytic inflammation with scattered plasma cells and CD20 reactive T cells and B cells. Azathioprine 50 mg daily and prednisone 60 mg tapered to 10 mg daily were started.

One month later, MRI cervical spine showed a slightly progressive enhanced dural thickening of the craniocervical junction extending into the cervical spine and azathioprine dose increased to 50 mg twice daily.

4 months after, she was admitted to the hospital for stroke like symptoms and myelopathy. Repeat MRI brain revealed the tumor to be the same size as it was in April 2019. Azathioprine was discontinued and prednisone increased to 60mg daily.

One month after stopping Azathioprine, she started cyclophosphamide 1000 mg biweekly. In January 2020, MRI brain & C-spine respectively, showed significant improvement of about 50% in dural thickening to the posterior fossa and upper cervical cord (see Figure 1B and Figure 2B).

Exactly one year later in early 2020, brain MRI showed no further improvement and Cyclophosphamide discontinued. Methotrexate was considered as an alternative treatment option; however, the patient had elevation in liver enzymes. Alternatively, rituximab was started at induction regimen of 375 mg per metered sq. infusions once weekly for 4 weeks. Post induction, she received two doses of 500 mg rituximab two weeks apart. Maintenance therapy plan was for 500 to 1000 mg infusions every 16 weeks.

The presence of a photosensitive malar rash with erythema, telangiectasias affecting the mentum, and persistence of skin lesions consisting of linear atrophic plaques with sparse subcutaneous calcified nodules and dimpling of the skin on the forehead, buttocks, thighs, back and arms did continue. The lesions were biopsied showing collagen vascular disease and interface dermatitis indicative of lupus erythematosus profundus (LEP). Dermatology suggested a deeper biopsy for further confirmation.

Images

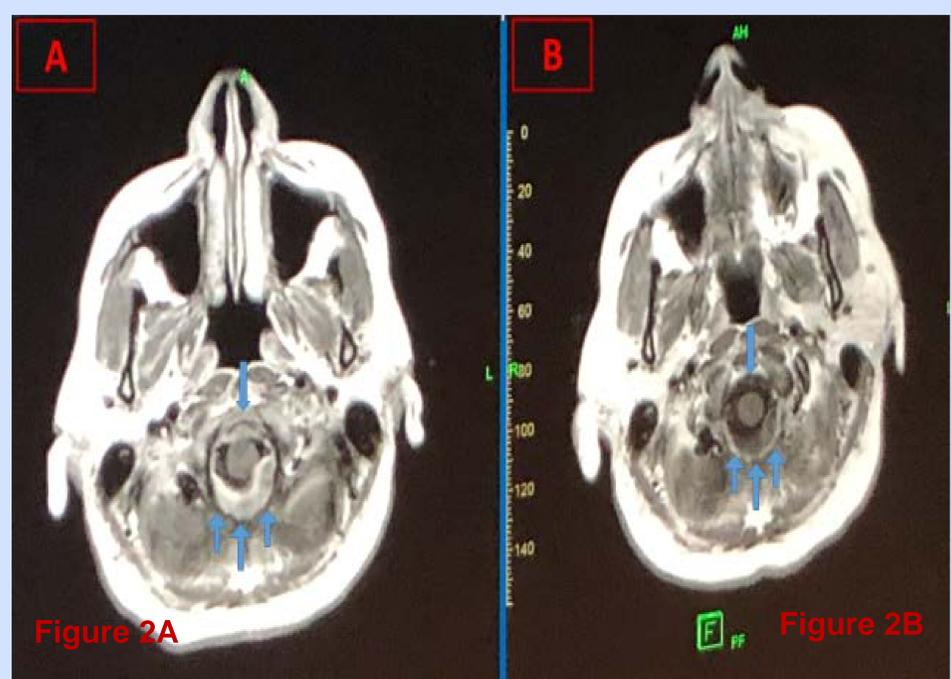
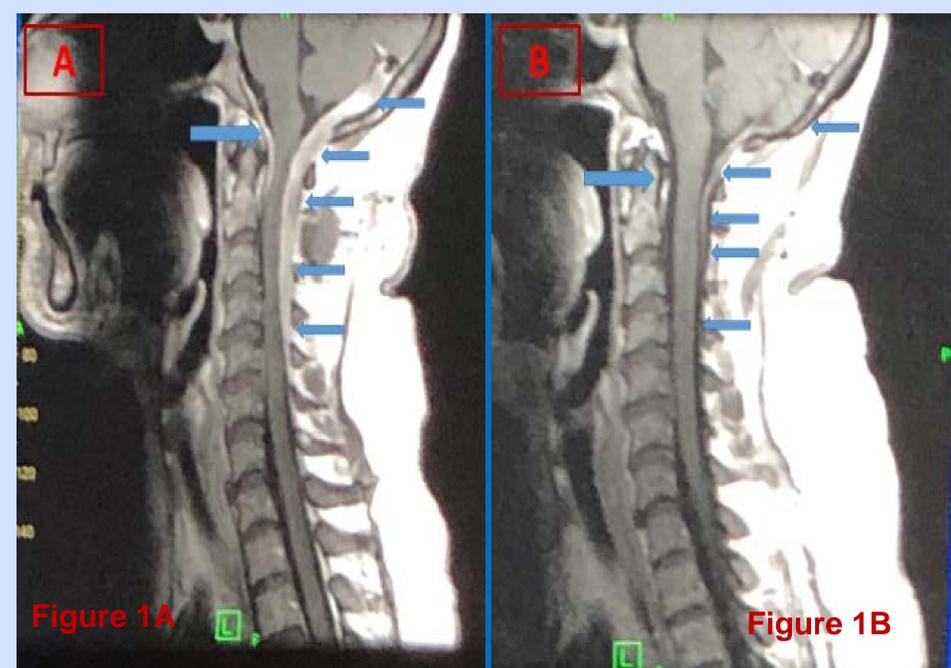


Figure 1A/2A Cervical spine MRI prior to treatment of cyclophosphamide showed an enhancing mass within the posterior fossa mass tracking inferiorly, with cervical extension, displaced to the upper cervical cord anteriorly causing significant impingement.

Figure 1B/2B Cervical spine MRI post treatment with cyclophosphamide showed significant 50% improvement in dural thickening to the posterior fossa and upper cervical cord. Measuring at 3 mm in thickness at the craniocervical junction (previously 6.2 mm).

Discussion

Vital in diagnosis of IHP, is contrast enhanced MRI [5, 6]. Biopsy, is the gold standard in the setting of progressive symptoms. Microscopic findings are usually significant for densely dispersed mixed inflammatory infiltrates, abundant lymphoplasmacytic cells, exuberant fibroplasia, and focal hyaline degeneration [7, 8]. These biopsy findings were evident in our patient.

It has been generally accepted that first line therapy is prednisone dosed at 1 mg / kg / day. Immunosuppressant medications are usually started if patients are refractory to the prednisone regimen, in which azathioprine, cyclophosphamide and methotrexate are most widely used [4, 9]. Our patient was initially started on prednisone. Azathioprine was added shortly after generating an initial response to treatment but, the patient later had a relapse of symptoms. Only after addition of cyclophosphamide was substantial improvement attained. Rituximab is suggested to be a good alternative therapy for steroid-refractory hypertrophic pachymeningitis (HP) and cases with idiopathic etiologies [11]. All patients should be followed using ESR, C-reactive protein, and MRI [4, 9].

From initial presentation in 2019, serial immunologic testing for antibodies remained completely negative or within normal limits.

Our patient had characteristic dermatologic findings suggestive of LEP. LEP is a form of cutaneous lupus erythematosus (CLE) characterized by chronic, recurrent subcutaneous inflammatory lesions appearing as erythematous, deeply indurated, tender plaques and nodules. CLE can present as distinct disease or in association with SLE and has a reported incidence of 4.3 per 100,000 [12]. Patients with CLE may never progress to SLE, yet according to population-based studies patients may demonstrate progression to SLE who initially had CLE ranging from 0-28%. Manifestation of cutaneous disease to SLE diagnosis can extended from months to 30 plus years [12-14]. CLE is typically divided into lupus-specific and nonspecific manifestations. Lupus-specific histopathology is distinct for lupus including vacuolar interface dermatitis [15]. Consistent with skin biopsy in our patient. Our patient had a presumptive diagnosis of cutaneous SLE (malar photosensitive rash, Raynaud's, oral sores and LEP). CLE i.e. LEP can develop into SLE, thus it is imperative to continue to follow rheumatological markers such as ANA and dsDNA for progression to SLE. A population based study evaluating, suggests regular lifelong screening [12].

HP many times idiopathic has been associated with various rheumatologic disorders most notably Rheumatoid arthritis, mixed connective tissue disease, and other vasculitides (SLE, Sjogren's and granulomatosis with polyangiitis). This case presents a probable association with CLE making it potentially the first reported case.

Conclusion

This report demonstrates a rare incidence of IHP in possible relation to CLE. It highlights the importance of pursuing suspicious findings on physical examination with proper imaging, lab analysis, and surgical intervention if necessary. Despite IHP being rare, all rheumatologic patients presenting with persistent headaches, neurologic signs, and now in conjunction with characteristic cutaneous features, should raise suspicion for further investigation.

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This research was supported (in whole or in part) by HCA Healthcare and/or an HCA affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.