Rare Case Of Plasma Cell Hyperplasia (Inflammatory Pseudotumor) Of The Central Nervous System

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Citation

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Abstract

We report a rare case of Plasma cell hyperplasia (PCH) of the Central Nervous System (CNS) in a 25 year old male involving the pachymeninges and brain parenchyma which was a diagnostic dilemma due to overlapping imaging features with other pathologies. On CT, there was white matter edema and gyral swelling in left parietoccipital region. The involved region showed ill-defined diffuse sheet like peripheral enhancement on post contrast CT, which was thought to be intraparencymal than pachymeningeal. On post contrast MR imaging, there was thick sheet like pachymeningeal enhancement with dural tail formation in addition to diffuse peripheral gyral enhancement and white matter edema in above location. Gross total excision of dural based lesion was done and histopathology revealed features of plasma cell hyperplasia. No evidence of recurrence was seen at six month follow-up.

INTRODUCTION

Plasma cell hyperplasia (PCH) also known as plasma cell granuloma (PCG), or inflammatory pseudotumor is a rare lesion. It was termed as inflammatory pseudotumor by Umikar et al in 1954 as it is an inflammatory process that can clinicoradiologically simulate a mass lesion.[1] Histologically, it is composed of a variable inflammatory and mesenchymal cellular mixture including plasma cells, histiocytes, lymphocytes and spindle cells. [2] Brain plasma cell granulomas are known to arise from the meninges with or without parencymal involvement, sella, choroid plexus, fourth ventricle, and hypothalamic region. [3,4,5]We report imaging features of this rare lesion involving both pachymeninges and brain parenchyma in a young male which was a diagnostic dilemma due to its overlapping features with other pathologies.

CASE REPORT

A 25 year old male presented with two month history of headache, vertigo, low grade fever and seizures. Gait was unsteady and patient had difficulty in passing stools.

CT revealed white matter edema and gyral swelling in left parietoccipital region. On post contrast study, there was ill defined diffuse peripheral enhancement in involved region. The enhancement on CECT was thought to be gyral within peripheral brain parencyma. Pachymeningeal involvement could not be separately picked up on CECT (Figure 1). Thus, on the basis of CT imaging features, possibilities of gliosarcoma and encephalitis were kept.

Subsequent contrast enhanced MRI revealed thick sheet like pachymeningeal enhancement in the parietoccipital location with dural tail formation and contiguous involvement of underlying brain which showed gyral swelling, diffuse peripheral gyral enhancement and white matter edema (Figure 2) .

MR features favoured possibilities of aggressive meningioma, tuberculosis and lymphoma. Aggressive meningioma was considered as most likely possibility due to localized nature of disease.

The patient underwent gross total excision of dural based lesion. Histopatholgical examination revealed a cellular lesion involving pachymeninges and brain parenchyma comprising lymphoplasmacytic infiltrate, proliferating fibrous tissue and glial elements with whorl formation, perivascular cuffing and dense chronic infiltrate in Virchow-Robin spaces. Findings were suggestive of plasma cell hyperplasia (inflammatory pseudotumor) of brain and pachymeninges (Figure3).

Post operative course was uneventful; the patient was discharged on steroids, mannitol and antiepileptic drugs. On six month follow up, he had persistant headache of reduced severity with visual field defects. Follow up CT, (Figure 4)

revealed encephalomalacia in left parietoccipital region with no recurrence.

{image:1}

{image:2}

{image:3}

Inset: Numerous plasma cells (blue arrow heads) with eosinophilic cytoplasm & nuclei pushed to the periphery.

{image:4}

DISCUSSION

Plasma cell hyperplasia of CNS is a rare inflammatory lesion of unknown etiology which can clinically mimic a neoplasm[1]. Histologically, it is composed of a variable inflammatory and mesenchymal cellular mixture including plasma cells, histiocytes, lymphocytes and spindle cells. Therefore, depending on the predominant cellular components, many synonyms for this disease have been described. It is also known as Inflammatory myofibroblastic tumor, inflammatory pseudotumor, xanthogranuloma and fibrous histiocytoma.[2] It is frequently reported in lung but other organs like CNS, liver, spleen, stomach, thyroid, kidney, bladder and orbits can be involved[6] .Majority of patients are under the age of 40 years and present with clinical features of mass lesion like headache, weakness, seizures, gait disturbances or visual deficits.[7, 8] Imaging shows involvement of brain, pachymeninges, orbits and nerves with diffuse sheet like pachymeningeal enhancement with or without underlying parenchymal involvement. Differential diagnosis on imaging is with aggressive meningioma, metastasis, lymphoma, tuberculosis, sarcoidosis, infiltrating neoplasms, xanthogranuloma [5, 8, 9, 10] and encephalitis with the closest differential being an aggressive meningioma because of its many overlapping features.

Thus, PCH can be difficult to diagnose only on the basis of imaging alone but may be considered as a diagnostic possibility if there is contiguous sheet like involvement of brain and pachymeninges in a young patient. Although imaging features can help, final diagnosis is by typical histopathological features. Histologically, it is characterised by dense lymphoplasamacytic infilterate involving brain and pachymeninges with scattered neutrophils.

Treatment is by surgical excision, steroids, radiation, antiepileptics and chemotherapy if needed. Long term follow up is required to detect recurrence.[8]

CONCLUSION

PCH is a rare and uncommonly diagnosed lesion. It has overlapping imaging features with various etiologies and may be kept as a diagnostic possibility on imaging in young patients with contiguous parencymal and pachymeningeal involvement since it has therapeutic implications.

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