CSF leak associated with POEMS syndrome

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Abstract

A 58-year-old Jamaican female presented with new onset diagnosis of papilledema **(Image set 1).** She had a prior history of an idiopathic peripheral neuropathy diagnosed as chronic inflammatory demyelinating polyneuropathy (CIDP) on intravenous immunoglobulin without improvement. In review of her history, she did have a **high monoclonal gammopathy, high serum platelet count, and darker skin changes**. The clinical history, lab testing and examination were suspicious for **POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, skin changes).**

A magnetic resonance imaging (MRI) was done which showed a **large empty sella and a left ethmoid sinus lesion which extended across the cribriform plate into the left anterior cranial fossa which was suspicious for a mass (Image set 2)**. Sinus endoscopy revealed a non-pulsating, reddish mass with some exudate over it in the left olfactory cleft area. A computer tomography (CT) scan done shortly after the procedure confirmed an **encephalocele (Image3)**. The patient was already on acetazolamide 500mg twice daily and undergoing treatment with bortezomib (Velcade) for the POEMS syndrome with a hematologist. The plan was to observe the skull base defect since it was asymptomatic.

18 months since the initial diagnosis of POEMS, she developed CSF leak from her nose. She underwent at trans-facial extradural resection of skull base meningoencephalocele with Alloderm closure of skull base defect/repair of CSF leak. She had a lumbar drain done prior to the surgery which measured the **opening pressure at 32cmH2O**.

42 months later, she had recurrence of the CSF leak. Repeat CT (Image 4) and MRI showed a larger empty sella and now a right sided meningoencephalocele. Repeat eye examination showed resolution of the initial papilledema. She underwent another repair of the CSF leak with findings this time showing a large meningoencephalocele reaching right nasal floor. Left sided repair previously done looked intact.

4 years after the second CSF leak repair, she remains without leak. The most recent MRI did not show any anterior skull base defects.

POEMS syndrome can present with papilledema due to intracranial hypertension as a result of high CSF protein.[1] CSF leak can be a complication of the chronic high cranial pressure.[2] **We saw no prior cases of POEMS syndrome and CSF leak rendering this case unique.** One case reported using ventriculoperitoneal shunt for symptoms and signs of high cranial pressure in POEMS.[3] We must consider POEMS syndrome in cases of high cranial pressure and other signs and symptoms atypical of classic idiopathic intracranial hypertension.

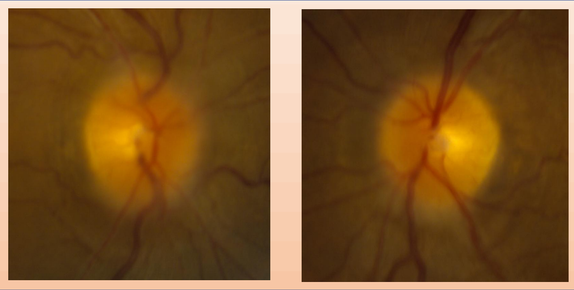
Introduction

POEMS syndrome is a rare disorder of **plasma cell origin**. POEMS syndrome stands for **polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes.[4]** Not all findings are seen in cases of POEMS syndrome and rather patients need to meet 2 mandatory, 1 major and 1 minor criteria for the diagnosis. **Elevated vascular endothelial growth factor (VEGF)** expressed by **plasma cells** may be the leading pathogenesis resulting in multisystem disease.[4] Papilledema (minor criteria) can occur in cases of POEMS syndrome due increased intracranial pressure (ICP).[5] Some cases of papilledema do not have high ICP and the mechanism of disc edema may be more inflammatory rather than due to abnormal CSF dynamics. CSF leak can be a complication of intracranial hypertension but there has never been a case of POEMS and CSF leak.

Methods and Materials

Single case report of a 58-year-old female with POEMS syndrome, papilledema and CSF leak due to recurrent encephalocele treated with surgery, acetazolamide and bortezomib (Velcade). MRI and CT images included showing pertinent findings.

Results



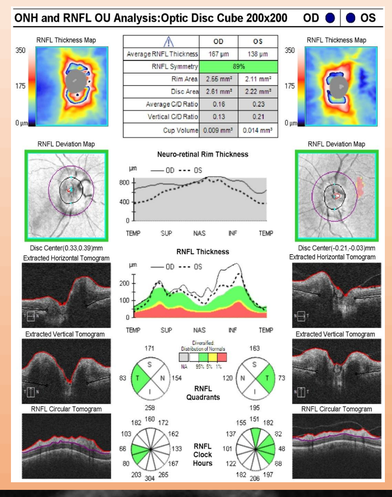


Image set 1. Optic nerve photos (top) showing mild bilateral disc edema and optical coherence tomography (OCT, right) showing mild thickening of the nerve fiver layer.

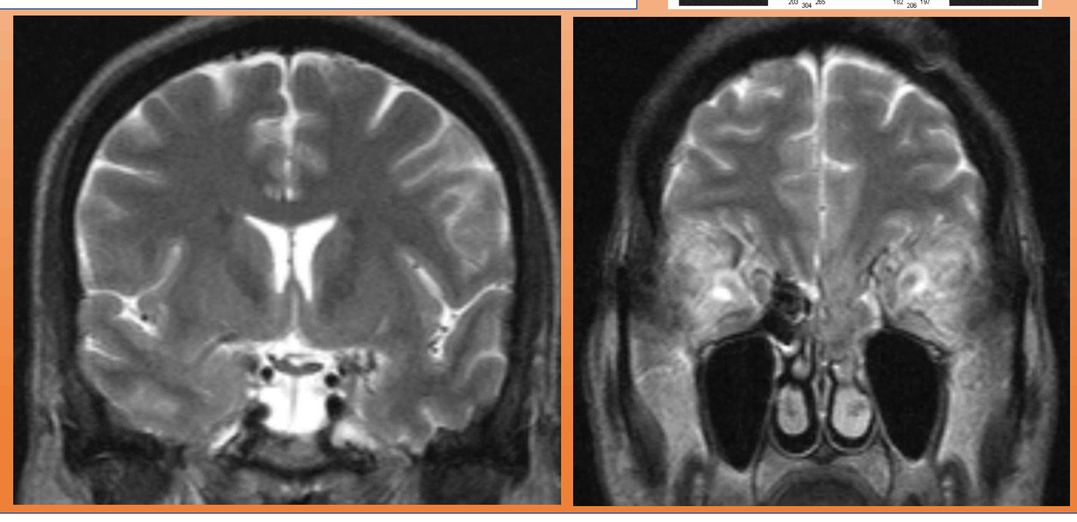
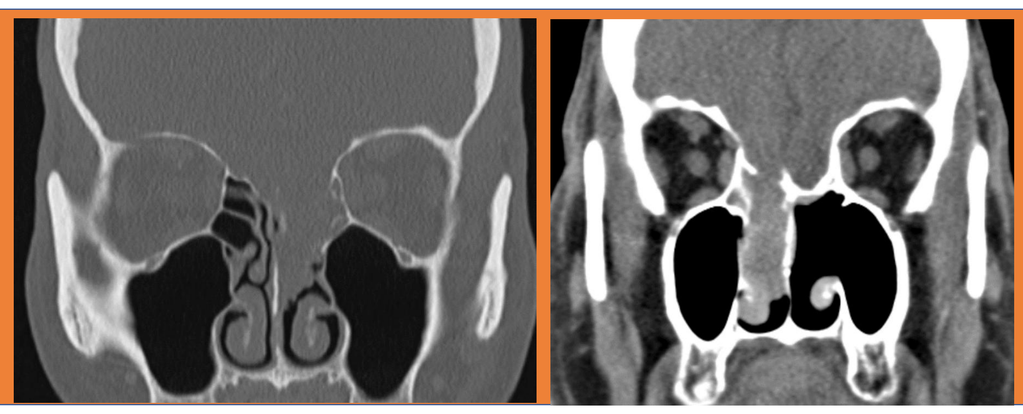


Image set 2. Coronal T2 magnetic resonance imaging (MRI) identifying a large empty sella (left) and a left ethmoid sinus lesion extending across the cribriform plate into the anterior cranial fossa (right).



Images 3 and 4. Initial coronal computer tomography bone window showing the left anterior skull base defect and the encephalocele. Follow-up coronal CT showing new right sided meningoencephalocele and prior treated left sided skull base 42 months later.

Discussion

Cerebral spinal fluid leak is a known complication of idiopathic intracranial hypertension (IIH).[2] Classic IIH symptoms are headaches, typically more when laying down, pulsatile tinnitus characterized as “whooshing” noise and transient visual alterations.[6] Most cases of IIH have papilledema. In cases of POEMS syndrome, papilledema is considered a minor criteria for the diagnosis.[4] Papilledema can occur in cases of POEMS due high ICP due high CSF protein. Most cases of papilledema associated with POEMS respond to acetazolamide and treating the underlying POEMS syndrome with medications targeting plasma cell proliferation. In this case, recurrent CSF leak occurred due to anterior skull base defects from high ICP. There have been no other cases of CSF leak in cases of POEMS syndrome although other cases of POEMS syndrome with high ICP have been treated with ventriculoperitoneal shunt.[3]

Conclusions

- CSF leak can be a complication of increased intracranial pressure.
- Papilledema is seen in most cases of intracranial hypertension.
- POEMS syndrome is a rare disorder of plasma cell origin that leads to multisystem inflammation characterized by high VEGF.
- Papilledema is a minor criteria in POEMS syndrome which can be due to high ICP.
- Cases of papilledema and POEMS respond to treating the plasma cell proliferation and acetazolamide.
- Rarely, neurosurgical intervention is needed for high ICP in POEMS.
- CSF leak in POEMS syndrome has not been reported.

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