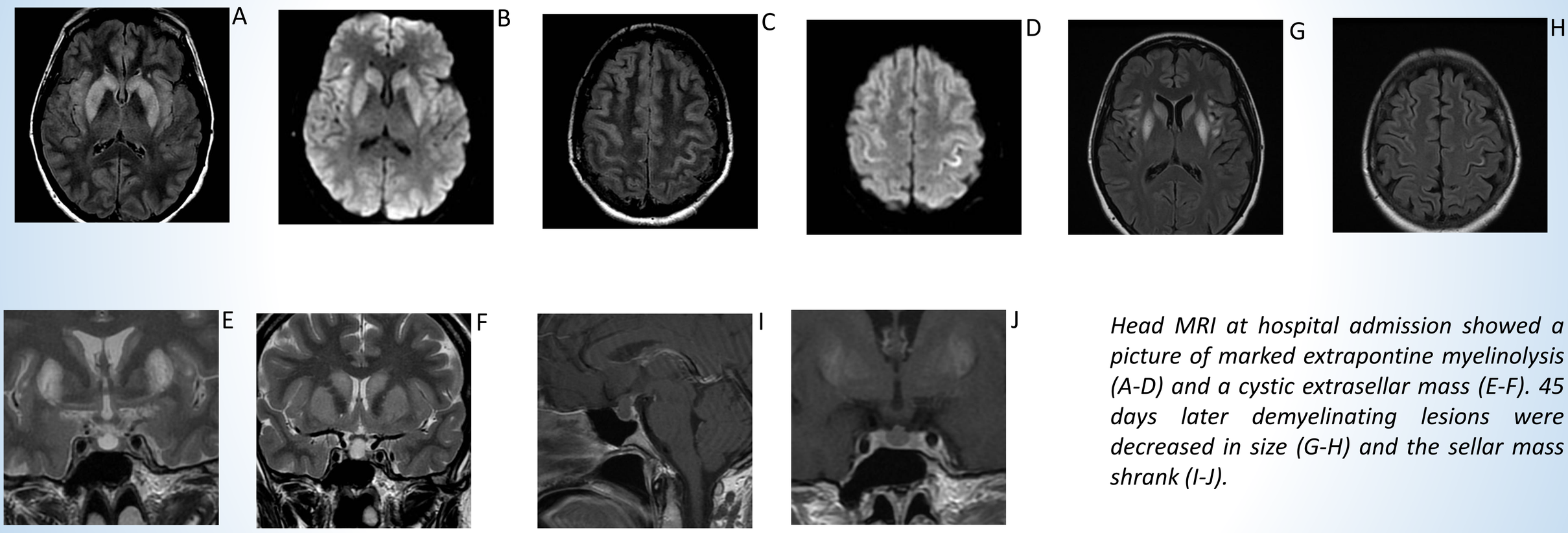
Extra-pontine myelinolysis showing good response to plasma exchange.

Oppo V[1], Brioschi M[1], Cozzi R[2], Doneda P[3], Erminio C[3], Gatti A[1], Jann S[1], Regna C[3], Verrengia E[1], Agostoni E[1].
1) Neuroscience Department, Niguarda Hospital, Milan
2) Endocrinology Department, Niguarda Hospital, Milan
3) Neuroradiology Service, Niguarda Hospital, Milan

Background: Osmotic Demyelination Syndrome (ODS) is a rare neurologic disorder that primarily occurs after rapid correction of severe hyponatremia. Pontine demyelination is the hallmark of the disease but solely extra-pontine lesions are seen in 12% of cases[1]. No specific treatment has proven efficacy.
Materials and methods: Case report and review of the literature.
Results: A 40-years aged woman, previously healthy, was referred to our neurology department because of an alterated behaviour and generalized seizures after correction of a profound hyponatremia (105 mEq/l). Neuroimaging showed a cystic sellar mass (most probably a Rathke's cleft cyst) and a picture suggestive of extra-pontine myelinolysis. She had an hypophysis function disorder with central hypothyroidism, hyperprolactinemia, hypogonadism and diabetes insipidus. Despite initial treatment with high-dose corticosteroids her neurological status worsened, she developed dysarthria, disfagia, bilateral facial palsy and moderate quadriparesis. She was then treated with 4 sessions of plasma exchange (PE) with a gradual marked improvement of neurological deficits. MRI at 1 and 3 month showed partial regression of demyelination and shrinkage of the sellar mass. 2 months after discharge the patient developed focal seizures with good response to antiepileptic therapy.



Head MRI at hospital admission showed a picture of marked extrapontine myelinolysis (A-D) and a cystic extrasellar mass (E-F). 45 days later demyelinating lesions were decreased in size (G-H) and the sellar mass shrank (I-J).

Discussion: In our case ODS followed correction of hyponatremia that likely resulted from a syndrome of inappropriate antidiuretic hormone secretion. Later in the clinical course she developed diabetes insipidus together with hyperprolactinemia, hypogonadism and central hypothyroidism. Hypophysis dysfunction took place probably because of a ruptured Rathke's cleft cyst. This relatively common benign lesion can be symptomatic for hypopituitarism and some reports exist of hypophysitis triggered by disruption of the cyst's wall and leakage of its high protein content[2]. Our patient markedly improved after PE, thus suggesting a beneficial effect of the procedure. Some reports describe a good response of ODS to PE, on the basis of removal of undefined myelinotoxic compounds released by osmotic stress[3]. On the other hand, spontaneous recovery with favourable outcome (described in 50% of cases)[1] cannot be excluded.
Conclusions: Effective therapies for ODS by now are lacking. PE is a feasible, promising therapeutic option that should be kept in mind by clinicians.

Bibliography

1) Singh TD, Fugate JE, Rabinstein AA. Central pontine ed extrapontine myelinolysis: a systematic review. Eur J Neurol 2014; 21: 1443-1450.
2) Schittenhelm J, Beschorner R, Psaras T et al. Rathke's cleft rupture as a potential initial event of a secondary perifocal lymphocytic hypophysitis: proposal of an unusual pathogenic events and review of the literature. Neurosurg Rev 2008; 31: 157-163.
3) Bibl D, Lampl C, Gabriel C et al. Treatment of central pontine myelinolyisis with therapeutic plasmapheresis. Lancet 1999 3;353(9159):1155.