Türe U et al. Hypothalamitis: A Novel Autoimmune Endocrine Disease. A Literature Review and Case Report. J Clin Endocrinol Metab. 2020 Oct 26:dgaa771. doi: 10.1210/clinem/dgaa771.

The relationship between the endocrine system and autoimmunity has been recognized for a long time and one of the best examples of autoimmune endocrine disease is autoimmune hypophysitis. A better understanding of autoimmune mechanisms and radiological, biochemical, and immunological developments have given rise to the definition of new autoimmune disorders including autoimmunity-related hypothalamic-pituitary disorders. However, whether hypothalamitis may occur as a distinct entity, is still matter of debate. Here we describe a 35-year-old woman with growing suprasellar mass, partial empty sella, central diabetes insipidus, hypopituitarism, and hyperprolactinemia. Histopathologic examination of surgically removed suprasellar mass revealed lymphocytic infiltrate suggestive of an autoimmune disease with hypothalamic involvement. The presence of anti-hypothalamus antibodies to AVP-secreting cells (AVPcAb) at high titers and the absence of anti-pituitary antibodies suggested the diagnosis of isolated hypothalamitis. Some similar conditions have sometimes been reported in the literature but the simultaneous double finding of lymphocytic infiltrate and the presence of AVPcAb so far has never been reported. We think that the hypothalamitis can be considered a new isolated autoimmune disease affecting the hypothalamus while the lymphocytic infundibulo-neurohypophysitis can be a consequence of hypothalamitis with subsequent autoimmune involvement of the pituitary. To our knowledge this is the first observation of autoimmune hypothalamic involvement with central diabetes insipidus, partial empty sella, anti-hypothalamic antibodies and hypopituitarism.