AN UNUSUAL CASE OF A PARA-SELLAR MASS

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Background

Pituitary lymphoma is very rare, although lymphomatous (or leukaemic) infiltration of the perisellar dura is not infrequently encountered as part of more widespread CNS disease. As is the case with CNS lymphoma elsewhere, pituitary lymphomas can either be primary (no systemic disease) or secondary (involvement of the pituitary gland is part of systemic disease). Pituitary lymphoma (i.e. parenchymal disease limited to the pituitary gland rather than dural involvement, or extension of cerebral primary CNS lymphoma to the region) is very rare, accounting for 0.1-0.3% of pituitary tumours. True primary lymphoma of the CNS is even less common, with only a smattering of cases reported.

Case Description

A 36 year-old female presented with increasing headaches over the previous 8 months. She was treated for migraines and cluster headache. She also had a history of depression. She had prodromal aura of visual disturbance including blurred vision and photophobia. She was admitted following sudden onset of stabbing pain over the left eye along with nausea. All these symptoms were classic of her usual migraines. On admission to hospital there was no focal neurological deficit but she was noted to have palpable lymph nodes in her right groin and neck. A full body CT scan revealed widespread enlargement of lymph glands, enlarged spleen, along with an abnormal enhancing focus with soft tissue nodularity in the para-sellar and suprasellar region. MRI of brain showed enlargement of the pituitary gland with displacement of the pituitary stalk. There was also an abnormal soft tissue focus in the right para-sellar region encroaching upon the sella. There was no evidence of involvement elsewhere in the brain. Baseline and dynamic pituitary function testing was normal.

Lymph node excision biopsy confirmed Hodgkin’s lymphoma of the nodular sclerosing type. Lumbar puncture was normal and bone marrow biopsy and trephine showed no evidence of lymphoma. She was diagnosed with stage IIIb Hodgkin's lymphoma. The nature of the para-sellar mass was uncertain as cranial involvement in Hodgkin’s lymphoma usually occurs in the terminal phase of the disease. The patient underwent chemotherapy with the ABVD regimen. A repeat MRI of the brain and pituitary after 2-months of chemotherapy showed a dramatic change with almost complete resolution of the para-sellar mass and normalization of the pituitary size. Repeated interval MRIs have been normal and the patient has remained in remission from her Hodgkin’s Lymphoma. This case illustrates an unusual presentation of a para-sellar mass due to Hodgkin’s lymphoma and its response to chemotherapy.

Investigations

MRI head pre-chemotherapy

MRI head post-chemotherapy

Discussion

Pituitary lymphomas are exceedingly rare in immunocompetent patients, exhibit variable clinical signs and symptoms, and carry poor prognosis. These tumours may represent metastasis of a systemic lymphoma, or a primary pituitary lymphoma. Systemic lymphomas may cause secondary CNS involvement but there are only few reported cases of pituitary involvement as the first presentation of these tumours especially in the absence of hypopituitarism and negative CSF analysis, as in our case.

Conclusion

In conclusion, we describe a rare case of parasellar mass, that present with diagnostic dilemma. These unusual cases emphasize the need for clinical awareness in such perplexing cases which clearly require a multidisciplinary approach.