SHORT REPORT

Sinus histiocytosis with massive lymphadenopathy—isolated suprasellar involvement

M B Bhattacharjee, S J Wroe, B N Harding, M Powell

Abstract

An unusual case of an isolated histioproliferative lesion arising from the suprasellar region is described. The presence of lymphophagocytosis suggested that this represented an extranodal intracranial form of sinus histiocytosis with massive lymphadenopathy.

Sinus histiocytosis with massive lymphadenopathy (SHML, Rosai-Dorfman disease), an uncommon histioproliferative disorder, presents typically with massive painless lymphadenopathy particularly in the neck, associated with fever, leucocytosis, elevated ESR, and hypergammaglobulinemia. Since its initial description in 1969¹ almost 400 cases have been collected in the SHML registry, and it has become an established clinicopathological entity. In over a quarter of cases the disease is extranodal² but usually the presence of massive lymphadenopathy facilitates diagnosis. Extranodal disease, however, may represent the predominant or

even exclusive manifestation. We report a case of isolated suprasellar involvement in a 78 year old man.

Case report

A 78 year old retired Welsh farmer presented in June 1989 with bilateral visual impairment. Eight months earlier he first noticed difficulty reading with the right eye and mild headache. The headache improved but his vision deteriorated progressively, and nine weeks before admission he noticed blurring of objects seen in the left side of the left visual field. Eventually he could see only hand movements with the right eye. He experienced no pain. He had no other symptoms, no important previous illnesses, did not smoke or drink, and had taken no drugs. There was no family history of neurological disease.

Examination showed an alert right handed man. Vision in his right eye was limited to counting fingers at 1 m. Visual acuity in the left eye corrected to 6/60, N12 with 11/13 Ishihara colour plates correctly identified. He had a bitemporal hemianopia, right optic atrophy, and right relative afferent pupillary defect. Other cranial nerves and neurological and general examination were otherwise normal. CT head scan showed an enhancing mass involving the sella (figure 1) extending superiorly with elevation of the optic chiasm and anteriorly to the planum sphenoidale. A delayed VER of low amplitude was recorded from the left whole and nasal half fields only. Pattern ERG was normal. Chest and skull x ray pictures, full blood count, ESR, and other routine investigations were all normal.

A granular tumour mass involving both optic nerves was subtotally removed through a right frontal craniotomy. He made a good postoperative recovery, and at eight days visual acuities corrected to 6/36, N35, and 3/13 colour plates on the right; 6/12, N10, and 13/13 colour plates on the left. His vision continued to improve, and at review in January 1990 visual acuities corrected to 6/12, N8, and 6/13 colour plates on the right; 6/9, N5, and 13/13 colour plates on the left. He had a right temporal hemianopia and right relative afferent pupillary defect. CT scan showed no suprasellar tumour recurrence. Small amplitude delayed VERs were recorded from the right eye with an increase in amplitude and shortening of latencies from the left eye.

National Hospital for
Neurology and
Neurosurgery and
Institute of Neurology,
Queen Square,
London WC1N 3BG

M B Bhattacharjee
S J Wroe
B N Harding
M Powell

Correspondence to:
Dr Harding

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Figure 1 Axial CT scans with contrast enhancement showing suprasellar extent of abnormal tissue with anterior extension along planum sphenoidale.

Figure 2 (a) Diffuse infiltrate of histiocytes, plasma cells, and histiocytes with lympho-phagocytosis highlighted in inset (H and E $\times 300$, inset $\times 460$). (b) Strong S-100 reactivity of histiocytes (avidin-biotin peroxidase method $\times 480$).

Pathological examination

Two fragments of pale tissue each measuring about 1 cm³ were processed in paraffin, and 5 μ m sections were stained with standard methods including those for demonstrating micro-organisms. Immunohistochemistry with antibodies to LCA (leucocyte common antigen), L26 (B cells), UCHL1 (T cells), S-100 protein, α -1-antichymotrypsin (ACT), lysozyme, and Kappa and Lambda light chains was performed with the avidin biotin conjugate method. Microscopic examination showed fibrous tissue with a diffuse infiltrate of histiocytes, plasma cells, and lymphocytes (figure 2a). The histiocytes had abundant pale pink cytoplasm and vesicular nuclei. Many of them showed within their cytoplasm numerous intact lymphocytes, a feature which has been designated as emperipolesis or lymphophagocytosis (figure 2a inset). There were small lymphoid aggregates and some plasma cells showing Russell bodies. There were no granulomas and no evidence of micro-organisms. Immunohistochemically the histiocytes were shown to be strongly reactive for S-100 protein (figure 2b) and ACT, the plasma cells to be polyclonal, and the lymphocytes present to be an admixture of B and T cells.

Discussion

Although extranodal pathology is seen in up to a third of cases of SHML,³ intracranial involvement is uncommon. Thirteen cases with CNS involvement have been reported^{4–10} including one of isolated dural disease causing trigeminal and facial nerve disturbance.¹⁰ In only one other case was there a suprasellar mass: a 32 year old man with a 10 year history of typical generalised SHML (cervical adenopathy, parotid and posterior mediastinal masses).⁹ In comparison to the nodal lesion, extranodal SHML is less easily recognised because the characteristic sinusoidal distribution of the histiocytes is lacking and lymphophagocytosis is less pronounced. Histiocytes in SHML share some of the markers of ordinary histiocytes but differ in consistently expressing S-100 protein.¹¹ SHML must be distinguished from Langerhan's cell histiocytosis (Histiocytosis X) as rare cases are localised entirely or predominantly within the CNS¹²; however these cells have a characteristic morphology as well as an associated eosinophil infiltrate. Although Langerhan's cells express S-100 protein, they lack phagocytosis and ultrastructurally show the Birbeck granule.¹³ The cells in malignant histiocytosis show clear atypia and necrosis and exhibit erythro- not lympho-phagocytosis.¹⁴

SHML usually affects people in the first two decades but no age group is exempt. The disease generally follows a protracted indolent course with eventual resolution in many cases,³ although a fatal outcome directly attributable to the disease has been reported in a few cases.^{15 16} Our patient was remarkable with respect to age and clinical presentation and unique in presenting as an isolated suprasellar mass involving both optic nerves. Of the various treatments attempted, only local surgical excision has been consistently successful. Surgical excision resulted in significant clinical improvement which was maintained at the one year follow up visit.

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- 1 Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy—a newly recognized benign clinicopathological entity. *Arch Path* 1969;87:63–70.
- 2 Foucar E, Dorfman R, Rosai J. Sinus histiocytosis with massive lymphadenopathy. Current status and future directions. *Arch Dermatol* 1988;124:1211–4.
- 3 Sanchez R, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. An analysis of 113 cases with special emphasis on its extranodal manifestations (abstract). *Lab Invest* 1977;36:21–2.
- 4 Song SK, Schwartz IS, Strauchen JA, et al. Meningeal nodules with features of sinus histiocytosis with massive lymphadenopathy. *Am J Surg Pathol* 1989;13(5):406–12.
- 5 Carey MP, Case CP. Sinus histiocytosis with massive lymphadenopathy presenting as a meningioma. *Neuropathol Appl Neurobiol* 1987;13:391–8.
- 6 Asai A, Matsutani M, Kohno T, et al. Leptomeningeal and orbital benign lymphophagocytic histiocytosis. Case report. *J Neurosurg* 1988;69(4):610–2.
- 7 Mir R, Aftalion B, Kahn LB. Sinus histiocytosis with massive lymphadenopathy and unusual extranodal manifestations. *Arch Pathol Lab Med* 1985;109(9):867–70.
- 8 Foucar E, Rosai J, Dorfman RF, Burnes RK. The neurologic manifestations of sinus histiocytosis with massive lymphadenopathy. *Neurology* 1982;32:365–71.
- 9 Friedman MJ, Rosoff LJ, Aftalion B, Khan A, Decker R, Steinberg H. Sinus histiocytosis presenting as a mediastinal mass. *Chest* 1984;86(2):266–7.

- 10 Trudel M. Dural involvement in sinus histiocytosis with massive lymphadenopathy. *J Neurosurg* 1984;60(4): 850-2.
- 11 Miettinen M, Paljakka P, Haveri P, Saxen E. Sinus histiocytosis with massive lymphadenopathy. A nodal and extranodal proliferation of S-100 protein positive histiocytes? *Am J Clin Pathol* 1987;88:270-7.
- 12 Kepes JJ, Kepes M. Predominantly cerebral forms of histiocytosis X. A reappraisal of "Gagels hypothalamic granuloma", "granuloma infiltrans of the hypothalamus" and "Ayala's disease" with a report of four cases. *Acta Neuropath* 1969;14:77-98.
- 13 Mieran GW, Favara BE, Brenman JM. Electron microscopy in Histiocytosis X. *Ultrastruct Pathol* 1982;3:137-42.
- 14 Warnke RA, Kim H, Dorfman RF. Malignant histiocytosis (histiocytic medullary reticulosis): I. clinicopathologic study of 29 cases. *Cancer* 1975;35:215-30.
- 15 Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. An analysis of 14 deaths occurring in a patient registry. *Cancer* 1984;54:1834-40.
- 16 Wright DH, Richards DB. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman's disease): report of a case with widespread nodal and extranodal dissemination. *Histopathology* 1981;5:697-709.