Relapsing intracranial Rosai-Dorfman disease

A Petzold, M Thom, M Powell, GT Plant

Abstract
Two patients presenting with recurrent visual impairment due to relapsing intracranial Rosai-Dorfman disease are described. In both patients a preoperative diagnosis of meningioma was made. Histological examination disclosed the characteristic picture of S100 and CD68 positive histiocytosis with prominent lymphophagocytosis. In both patients complete tumour removal by surgery was impossible with residual tissue being the origin of relapsing disease. Low dose radiation led to partial recovery of vision and resolution of the intracranial mass. Review of the literature on intracranial Rosai-Dorfman disease leads to the suggestion that postoperative radiotherapy may be advisable in all cases.

Keywords: Rosai-Dorfman; sinus histiocytosis; multiple meningioma; intracranial neoplasms

Rosai-Dorfman disease or sinus histiocytosis is a histiocytic proliferative disorder.1 Generally patients present in their mid-20s with cervical lymphadenopathy (87%), often preceded by a short non-specific infection.2 Extranodal involvement occurs in 25% to 43%2,4 and affects the skin (12%), paranasal sinuses (11%), soft tissue (9%), bone (9%), salivary gland (5%), oral cavity (3%), kidney (2%), lower respiratory tract (2%), larynx (1%), and, rarely, other locations.2 Intracranial lesions are extremely rare. To our knowledge 32 patients with intracranial masses have been described previously,3–30 including three with suprasellar and intrasellar locations.2 Intracranial lesions lead to the suggestion that postoperative radiotherapy may be advisable in all cases.

Patient 1
A 78 year old retired Welsh farmer presented at Moorfields Eye Hospital in June 1989 with progressive bilateral visual impairment. He underwent bifrontal craniotomy with subtotal tumour resection. His condition remained stable until January 1990.4 The patient presented, however, with further impaired vision in February 1990 and the MRI disclosed evidence of local recurrence of the mass around both optic nerves, which enhanced with gadolinium on T1 axial brain scans. His visual acuity had declined to 6/24, N24 and 2/13 Ishihara plates on the right; counting fingers at 1 m and only the Ishihara control plate on the left. In addition to the previous bitemporal hemianopia the left nasal field had decreased to a small “island of vision”. Both fundi showed loss of nerve fibre layer.

The patient underwent low dose radiotherapy, after which he showed improvement of his visual acuity to 6/18, 11/17 Ishihara plates on the right; 6/18, 16/17 Ishihara plates on the left. He remained clinically stable for the next 10 years. He had diabetes mellitus and a transient ischaemic attack in 1998. In 1999 he complained of some confusion and blurred vision. No further investigation was undertaken. He died of an unknown cause at the age of 89.

Patient 2
A 47 year old service engineer with rapidly developing right sided visual loss presented at the National Hospital for Neurology and Neurosurgery in July 1998. Six weeks previously he had complained of visual impairment and was found to have visual acuity of 6/24 on the right. Vision had decreased to light perception only 2 weeks later. On reflection the patient thought he had developed right sided visual loss. On examination he had left shoulder pain, weakness in his right leg, fatigue, and frequent headaches starting in the neck and radiating to the occiput. The medical history showed a lumbar disc prolapse 2 years previously.

On examination his visual acuity corrected to 6/6, N12 and 13/13 Ishihara charts correctly identified on the left. No light could be perceived on the right and there was a right afferent pupillary defect. Loss of nerve fibre layer was apparent on the right. Apart from a lateralised Weber test to the right with normal Rinne all other cranial nerves were normal. Sense of smell was not tested at the time. Sensory examination, tone, power and reflexes in the lower limbs were normal. No residual deficit from the previous disc prolapse could be shown. General examination was normal and there was no lymphadenopathy.
Brain CT at admission showed multiple mass lesions around the foramen magnum, in the chiasmatic cistern arising from the planum sphenoidale, above the cribriform plate, the right parafalcine region, and the cerebellopontine angle. These lesions were gadolinium enhancing on the MRI T1 images and suggestive of multiple intracranial neoplasms such as meningioma (fig 1 A). Chest radiography was normal for heart and lungs, but the left acromion showed a cystic lesion. No additional skeletal lesions were detected in whole body scintigraphy.

The laboratory investigation showed a decreased concentration of thyroid stimulating hormone (0.2 mU/l, normal range 0.25–5.0 mU/l) but normal T3 and T4; increased blood glucose (9.8 mmol/l, normal range 3.3–9.0 mmol/l), and increased white cell count of $14.5 \times 10^6/l$ with $12.8 \times 10^6/l$ neutrophils, $1.2 \times 10^6/l$ lymphocytes, $0.4 \times 10^6/l$ monocytes, and $0.1 \times 10^6/l$ eosinophils. The erythrocyte sedimentation rate was normal (9 mm/h).

He underwent subtotal transglabellar resection of the suprasellar mass. The postoperative visual fields showed a small “island of vision” on the right and a superior depressed field on the left. His visual acuity corrected to 6/5 on the left and to finger count on the right. The MRI 1 month after the operation showed some residual gadolinium enhancing tissue around the optic chiasm (fig 1 B).

One year later the patient complained of recurrence of the visual symptoms. His visual acuity worsened to 6/9 on the left and to light perception on the right. His sense of smell was now impaired. On MRI the sellar mass had increased in size and extended superiorly, causing bowing of the infundibulum and tilting the optic chiasm (fig 1 C). The patient underwent radiotherapy (20 Gy over 10 fractions). Three months later visual acuity had improved to counting fingers on the right and 6/5 on the left. His visual fields and colour vision were full on the left. On the right he could perceive hand movements in all quadrants apart from the lower temporal. At 1 year follow up the MRI showed decrease of the sellar mass (fig 1 D). Some residual tissue could still be seen on the axial images.

**Histology**

The histology of patient 1 has been discussed in detail by Bhattacharjee et al and has been reviewed by one coauthor (MT); it is identical to that of patient 2.

For patient 2 histology of the suprasellar dural mass (30×20×8 mm) showed collagenous tissue with mixed chronic inflammatory cells and numerous polymorphs with very occasional eosinophils. Interspersed within this infiltrate were large cells with vesicular nuclei, some with indented nuclear membranes and abundant cytoplasm. Prominent lymphophagocytosis (fig 2 A) and phagocytosis of neutrophils and red blood cells by these cells was noted. Immunohistochemistry with positive labelling for S100 (fig 2 B) and CD68 (PKM1) and negative staining for CD1a confirmed the histiocytic cell lineage.

**Discussion**

Rosai-Dorfman disease or “sinus histiocytosis with massive lymphadenopathy” was initially...
is clearly seen (haematoxylin and eosin, original magnification ×625). Lymphophagocytosis (empoiopolesis) by the histiocytic cells nuclei, some with nuclear indentations, are seen. There is an associated infiltrate of lymphocytes within the histiocytes. Because of the occurrence of extranodal disease in about 43% of patients the term “Rosai-Dorfman disease” is widely used in the literature for this subgroup.7

Figure 2  Histology from patient 2. (A) Sheets of histiocytic cells with large vesicular nuclei, some with nuclear indentations, are seen. There is an associated infiltrate of polymorphs and lymphocytes. Lymphophagocytosis (empoiopolesis) by the histiocytic cells is clearly seen (haematoxylin and eosin, original magnification ×375). (B) The histiocytic cells show strong nuclear and cytoplasmatic staining with S100 protein and lymphophagocytosis is also seen (S100 immunohistochemistry, original magnification ×625).

In summary it seems advisable to ensure a 5 year follow up period (median relapse time) including brain imaging in patients with intracranial Rosai-Dorfman disease. The main preoperative diagnosis remains meningioma, but histology should establish the diagnosis. In cases with subtotal tumour resection or recurrence of neurological symptoms we would treat with local low dose radiation early rather than late.

1 Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy; a newly recognized clinico-pathological entity. Arch Pathol (Chicago) 1969;82:63–70.

www.jmp.com
Relapsing intracranial Rosai-Dorfman disease


