Sarcoid related optochiasmatic arachnoiditis: favourable outcome confirmed with MRI

A 57 year old woman was admitted for bilateral visual loss, most marked on the right. Her medical history included longstanding pulmonary sarcoidosis (stage 1) with bilateral hilar adenopathy, which had been histologically established. No specific treatment had been given.

She presented with rapidly progressive visual loss over several days with pain on ocular movement and headache. The patient was asthenic, pale, and afebrile. The initial visual acuity was 1/6 on the right eye and 3/6 on the left eye. Ophthalmic examination showed bilateral pallor of the papillae but no uveitis. The rest of the clinical examination, including the respiratory and neurological systems, was unremarkable.

Laboratory examination disclosed an inflammatory syndrome with an erythrocyte sedimentation rate of 45 mm in the first hour. Examination of the CSF showed an aseptic meningitis with an increased protein concentration (81 cells/mm$^3$ (99% lymphocytes), protein 1.15 g/l without intrathecal IgG synthesis). The blood concentration of angiotensin converting enzyme was increased (0.36 nmol/min/ml (normal 0.06–0.20)). A radiograph of the chest and an EMG showed no abnormalities.

Cranial MRI (figure, top panels) showed hypertrophy of the optic chiasma and of the optic nerves, with hypersignal on T2 weighted images (A and C, black arrow) and (B and D, arrowhead) gadolinium enhancement on T1 weighted images. Images (E–H) after corticosteroid treatment showed a decrease in the size and the intensity of the signal of both the optic chiasma and nerves (E and G), with disappearance of gadolinium enhancement (F and H). A, B, E, and F are in the axial plane; C, D, G, and H are in the coronal plane; A, E, C, and G are T2 weighted images; B, F, D, and H are post-gadolinium, T1 weighted images.

The patient was treated with high doses of steroids—an infusion of 1g/day methylprednisolone for 3 days, followed by 1 mg/kg/day oral prednisone for 1 month. Her overall condition improved. Ocular pain resolved rapidly over 2–3 days and visual acuity returned to normal (visual acuity =5/6 on both sides) within 4 weeks. The laboratory indices of inflammation and the CSF returned to normal within 6 weeks.

A control MRI (E, F, G, and H) was performed 2 months after the onset of treatment: a decrease in the size of both the chiasma and optic nerves, as well as the hypersignal on T2 weighted images (E and G), was seen. The post-gadolinium enhancement on T1 weighted images disappeared (F and H).

Steroid treatment is effective in optochiasmatic arachnoiditis due to sarcoidosis$^{1–3}$ and the response to treatment can be easily monitored clinically and with cranial MRI.$^1$

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