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LETTER TO THE EDITOR

Cavernous sinus syndrome due to neurosarcoidosis in adolescence: a diagnosis not to be missed

F. Rosini[1] · D. Bennett[2] · A. Cerase[3] · L. Volterrani[4] · A. Federico[1] · P. Rottoli[2] · Alessandra Rufa[1]

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Dear Sir,

Cavernous sinus syndrome (CSS) is a pathological condition characterized by usually unilateral painful ophthalmoplegia associated with headache, diplopia, ptosis, pupillary changes, trigeminal nerve dysfunction and retro-orbital pain. Primitive or metastatic malignancies, vascular abnormalities and inflammatory processes are the most common causes of CSS [1]. Differential diagnosis between the various conditions may be tangled; in this respect, laboratory and brain MRI findings are discriminatory [2]. We describe here a rare case of unilateral CSS as the presenting sign of neurosarcoidosis in adolescence.

A 16-year-old female referred to our neuro-ophthalmological outpatient service for right ptosis, non-reacting midriasis, pain during eye movements, cephalalgia, and diplopia. She also referred mild exertional dyspnea and fatigue in the last months. Neurological and neuro-ophthalmological examinations showed complete right III cranial nerve palsy with non-reacting midriasis, congested right optic disc with a normal neurorim, in absence of other neurological defects. Visual field was normal, while visual evoked potential showed delayed latency of P100 in the right eye. Head MRI (Fig. 1) showed a pachymeningeal thickening of the right cavernous sinus region, extending toward ipsilateral orbital fissures and oval foramen, and an analogous focal pachymeningeal thickening in both the temporal regions. All these lesions showed gadolinium enhancement. Autoimmune screening, thyroid function, hematological testing and Mantoux reaction were normal. While serum ACE and lysozyme dosage were in the normal range, serum chitotriosidase was increased (64 nmol/h/ml, cut-off limit 48.8 nmol/h/ml) [3]. Cerebrospinal fluid (CSF) analysis and investigation for *Mycobacterium tuberculosis*, neurotropic viruses and culture for bacteria were negative. Pulmonary function tests (PFTs) were normal, but diffusing capacity of lung for carbon monoxide (DLCO) was reduced (57% of predicted). Chest high-resolution CT scan (HRCT) showed a mild parenchymal involvement with peripheral and perifissural micronodules suggestive of sarcoidosis. Bronchoalveolar lavage (BAL) revealed an increased percentage of lymphocytes with high CD4/CD8 ratio (macrophages 46.5%, lymphocytes 46.5%, neutrophils 3.5%, eosinophils 3%, basophils 0.5%; CD4/CD8 lymphocytes ratio 3.98). Therefore, diagnosis of pachymeningeal and lung sarcoidosis was considered consistent and pulsed intravenous corticosteroid therapy (1 g/day) for 5 days, followed by oral therapy, was introduced. The therapy achieved a rapid, marked and durable improvement of symptoms. Six-month follow-up showed resolution of all clinical aspects, with only remaining reacting-midriasis, complete disappearance of meningeal thickening at MRI and DLCO improvement (70% pred.). Steroid therapy was tapered till complete suspension 24 months

F. Rosini and D. Bennett equally contributed to the present paper.

✉ Alessandra Rufa
rufa@unisi.it

[1] Clinical Neurology and Neurometabolic Diseases Unit, Department of Medical, Surgical and Neurological Sciences, University of Siena, Viale Bracci 2, Policlinico Santa Maria alle Scotte, 53100 Siena, Italy

[2] Respiratory Diseases and Lung Transplant Unit, Department of Medical, Surgical and Neurological Sciences, University of Siena, Siena, Italy

[3] Neuroimaging and Neurointervention Unit, NINT, AOUSenese, Siena, Italy

[4] Department of Medical, Surgical and Neurological Sciences, Diagnostic Imaging, University of Siena, Siena, Italy