Cervical epidural pseudotumor and multifocal fibrosclerosis

Case report and review of the literature

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The authors present the case of a 45-year-old man suffering from progressive quadriplegia due to an expansive C3–T2 epidural mass. Neuropathological examination demonstrated pseudotumor tissue. The patient had had an orbital pseudotumor 5 years before admission, and other systemic manifestations of an idiopathic inflammatory disease were discovered. This case is extremely rare. Nine cases of multifocal fibrosclerosis with central nervous system involvement are described in the literature. To the authors’ knowledge, this is the first description of a cervical epidural pseudotumor. Modern imaging has made the diagnosis of such an entity possible, and it is important for the neurosurgeon to consider this syndrome because the combination of surgery and systemic medical therapy can ensure a long-term survival with good quality of life.

Key Words • pseudotumor • multifocal fibrosclerosis • epidural lesion • cervical spine • orbit

In 1905 Birch-Hirschfeld described the first patient with an orbital pseudotumor. Nearly a century later, many clinical features with similar histological characteristics have been reported in various anatomical locations. We present the first case, to our knowledge, of a cervical epidural pseudotumor in a patient with a proven history of multifocal fibrosclerosis manifested by a typical orbital pseudotumor and three other sites of similar problems: a perirenal fasciitis and bilateral pleural effusions. This case of multifocal fibrosclerosis will be discussed in the context of the present knowledge about this rare syndrome.

Case Report

History. In June 1987, this 45-year-old man presented for the first time with a progressive painful right-sided exophthalmos. A ptosis appeared within 1 month, and movement of the right eye became limited in the upward and right direction (Fig. 1 left). Physical examination revealed a trigeminal (first branch) hypesthesia as well as multiple bilateral cervical and axillary adenopathy. The results of laboratory testing revealed only an inflammatory syndrome (ESR 37 mm/hour; C-reactive protein 27 mg/L). Ophthalmological examination confirmed a normal fundus and normal visual acuity. A CT scan of the orbit revealed an infiltrative process (Fig. 1 center) in the upper part of the right orbit with upper muscle thickening. Examination of a cervical lymph node biopsy sample demonstrated an aspecific adenitis. Short-term high-dose corticosteroid therapy induced a regression of the exophthalmos, disappearance of the CT-documented features (Fig. 1 right), and a normalization of the biological disturbances. The efficacy of the corticosteroid therapy confirmed the diagnosis of an orbital pseudotumor.

Examination. In October 1992, the patient developed a bilateral cervicobrachial neuralgia with interscapular irradiation, without any typical radicular topography. The pain was most severe during the night, and no relief was obtained after initiating nonsteroidal antiinflammatory treatment. In November 1992 the patient was readmitted to our department because of the recent onset of a progressive quadriplegia with distal dysesthesias. Neurological examination suggested that moderate spinal cord compression was present at the C-7 level. The patient was ataxic and experienced abolition of the vibration sense, distal weakness, and bilateral pyramidal syndrome. Urinary retention and bowel incontinence were observed. The results of standard

Abbreviations used in this paper: CNS = central nervous system; CT = computerized tomography; ESR = erythrocyte sedimentation rate; MR = magnetic resonance.
cervical radiography were normal. Magnetic resonance imaging of the cervical spine (Fig. 2) revealed an epidural expansive process from C-3 to T-2.

**Operation and Pathological Examination.** A C3–T2 laminectomy was performed via a posterior surgical approach. We found a fibrous epidural lesion adherent to the dural plane. The dura mater was intact, and no thickening was observed. Histological examination (Fig. 3) of the epidural conjunctival tissue sample demonstrated a lymphoplasmocytic infiltration with follicular organization and the presence of eosinophilic cells. Immunohistochemical findings indicated a chronic inflammatory reaction rather than a lymphoma. No germs or foreign organisms were found in the samples.

**Postoperative Course.** Biological examination revealed a moderate inflammatory syndrome (ESR 33 mm/hour; C-reactive protein level 33 mg/L; and fibrinogen level 469 mg/dl). Thyroid function, rheumatoid factors, and serum levels were normal. Intradermal reaction was negative. Cerebrospinal fluid analysis demonstrated a pleocytosis with 64 cells/mm³, 82% neutrophils, and an increased protein level of 397 mg/dl. Ophthalmological examination demonstrated normal findings. Abdominal CT scanning revealed perirenal fascia thickening and liquid in the Douglas pouch. Thoracic CT scanning confirmed the presence of bilateral pleural effusions that were suggested by clinical examination.

**Follow-Up Course.** After initiation of corticosteroid therapy, the patient’s neurological status began to improve. Indeed, after 2 months, he was able to walk alone despite a slight ataxia caused by some persistent sensory deficits. Urinary function returned without additional treatment. Postoperative MR imaging (Fig. 4) did not demonstrate any residual tumor. According to the treatment for multifocal fibrosclerosis, a 32-mg/day dosage of methylprednisolone was continued. A concurrent psychiatric disease, not related to steroid therapy, was responsible for the patient’s sudden death by suicide, and pathoanatomical examination was not performed.

**Discussion**

In the evaluation of an epidural pseudotumor, an extensive differential diagnosis must be considered. An infection must be excluded. In our case, bacteriological cultures of all the biopsy samples were negative for infection. Serological testing was negative. Because of the paucity of eosinophils, a parasitic infection was unlikely. The presence of a lymphoma can also account for such a clinical presentation, but this lesion was excluded by immunohistochemical studies, which demonstrated a polyclonal infiltrative picture of various T and B lymphocytes. Granulomatosis diseases such as sarcoidosis and Wegener granulomatosis were ruled out because there were no morphological features of vasculitis or granuloma formation. After the exclusion of all other causes in our case, the diagnosis of an epidural pseudotumor was considered. This case is unique because a pseudotumor has never been reported in this anatomical location. The diagnosis of an orbital pseudotumor was confirmed by its favorable outcome.
Involvement of the CNS is extremely rare. We found nine additional cases of multifocal fibrosclerosis with CNS involvement published between 1969 and 1999 (Table 1). These included cases of a hypertrophic intracranial pachymeningitis with patchy (8–10-mm) thickening in the dural plane and, in the others, intra- or parasellar pseudotumors. Tanaka, et al., have described a dorsal epidural location. Our cervical epidural lesion was unique, because of a normal dura mater. On the other hand, intracranial extension of orbital pseudotumor occurring in the absence of systemic illness—suggestive of multifocal fibrosclerosis—has been reported in seven cases, but we found only two cases of isolated cervical pseudotumor.

It is important to distinguish the syndrome of multifocal fibrosclerosis from that of pachymeningitis hypertrophia, limited to the CNS, for which several causative factors have been recognized: traumatic injury, toxins, metabolic diseases, rheumatoid arthritis, syphilis, tuberculosis, fungal meningitis, or isolated intracranial pseudotumors.

This entity is best treated with corticosteroid therapy, the efficacy of which has been well established. The main goal of surgery is to obtain histological diagnosis, but in some cases, a decompressive procedure permits quick improvement; however, the disease may recur after surgical intervention unless it is accompanied by some form of systemic therapy. The value of chemotherapeutic agents, such as azathioprine and cyclophosphamide, is uncertain. Radiotherapy has also been used, although scientific evidence of its efficacy has not been published.
<table>
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<td>NS</td>
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<td>corticotherapy</td>
<td>benefit from op &amp; corticotherapy, death from suicide</td>
<td>2 yrs</td>
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* CRP = C-reactive protein; CSF = cerebrospinal fluid; hyperprot = hyperproteinemia; incr = increased; NS = not specified.  
† These two groups report on the same patient.
Conclusions

The orbital pseudotumor is well known by ophthalmologists. It is important for neurosurgeons to consider the existence of idiopathic inflammatory manifestations involving all parts of the CNS, sometimes with or without the associated systemic disease termed multifocal fibrosclerosis, especially in cranial base surgery. A careful review of patient history, physical examination, and observation should be sufficient for the recognition of the manifestations associated with this syndrome, but acute diagnosis is based on results obtained from histological examination. After diagnosis, steroid therapy is initiated as the treatment of choice. The place of surgery is limited to decompression when the pseudotumor process is responsible for rapid neurological deterioration.

References

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