Dural involvement in sinus histiocytosis with massive lymphadenopathy

Case report

MICHEL TRUDEL, M.D., F.R.C.P.(C)
Department of Pathology, Faculty of Medicine, McGill University, Montreal, Quebec, Canada

An unusual case of an isolated histioproliferative lesion arising from the basal dura of the left middle cranial fossa is described. The presence of lymphophagocytosis suggests that this represents an extra-nodal, intracranial form of sinus histiocytosis with massive lymphadenopathy.

KEY WORDS • sinus histiocytosis • lymphadenopathy • cranial dura • histiocytosis X

Since its initial description by Rosai and Dorfman in 1969, sinus histiocytosis with massive lymphadenopathy (SHML) has become an established clinicopathological entity. The major features include prominent cervical lymphadenopathy with fever and neutrophilic leukocytosis with raised erythrocyte sedimentation rate (ESR). The sinuses of involved lymph nodes are typically distended with well differentiated histiocytes, many of which show the distinctive phenomenon of lymphophagocytosis. The disease follows a protracted course, but complete recovery eventually occurs in the majority of cases.

Extra-nodal disease has been recorded in various sites, most commonly the orbit and eyelid, upper respiratory tract, and skin. Involvement of the central nervous system is rare. In two reported cases, spinal epidural deposits produced paraparesis. This article presents the clinical and pathological findings in the case of a histioproliferative lesion confined to the dura of the left middle cranial fossa. The microscopic appearance was that of SHML.

Case Report

This 28-year-old man presented to a general hospital with a 1-year history of left facial numbness and paresthesias, and left-sided hearing loss. Six months before admission, he experienced the onset of left-sided headaches which gradually increased in frequency and severity. He also complained of unsteadiness of gait. There were no other symptoms.

Examination. The patient was alert and cooperative. There was decreased sensation to pinprick on the left side of the face. A left-sided perceptive hearing deficit was confirmed by audiometry (80% loss). Other cranial nerves were normal. Tandem gait was impaired. General examination disclosed no cutaneous eruption, lymphadenopathy, or hepatosplenomegaly.

Routine hematological and biochemical studies were normal. Computerized tomography revealed a lesion involving the medial portion of the left petrous bone. Vertebral angiography demonstrated displacement of the left posterior cerebral artery. The preoperative diagnosis was meningioma.

Operation. At left subtemporal craniotomy, an ill-defined mass was found infiltrating the basal dura of the left middle cranial fossa. It eroded the underlying portion of the left petrous bone, and erupted through the floor of the left middle cranial fossa into the pterygoid fossa. The lesion extended anteriorly to the foramen ovale, laterally to the opening of the vestibular aqueduct, and posteriorly to the condylar canal. The trigeminal ganglion was displaced and compressed, and there was slight displacement of the brain stem to the right. Approximately 90% of the mass was removed piecemeal in small fragments.

Pathological Examination. Microscopic examination showed infiltration of the dura by a mixed cellular population composed predominantly of histiocytes, intermingled with lymphocytes and plasma cells. The infiltrate was set in a fibrocollagenous stroma. The
Intracranial sinus histiocytosis

FIG. 1. Computerized tomography scan showing a mass destroying the medial portion of the left petrous bone.

The histiocytes were mature, with one or more indented nuclei and abundant eosinophilic cytoplasm. In some areas, the histiocytes had foamy cytoplasm, imparting a xanthomatous appearance to the tissue. A striking feature was the presence of lymphocytes and occasional plasma cells within the cytoplasm of several histiocytes (Fig. 2).

Lymphocytes were scattered throughout the lesion, occasionally forming small lymphoid aggregates. Many plasma cells contained Russell bodies. Eosinophils were inconspicuous.

Special stains for bacteria, acid-fast bacilli, fungi, and parasites were negative. Electron microscopy demonstrated predominantly typical macrophages, some of which showed lymphophagocytosis. No Langerhans' granules were identified.

Postoperative Course. After surgery, the patient showed a transient upper motor neuron facial weakness on the right side. A neuroparalytic keratitis in the left eye resolved spontaneously. His headaches and unsteadiness ceased. Additional investigations, including serum immunoelectrophoresis, iliac crest marrow aspiration, and chest and skeletal x-ray films were negative. A 1-week course of local radiotherapy (1500 rads) was given. Left facial hypesthesia persisted for several months after surgery but gradually resolved. On follow-up examination 14 months after surgery, the patient was in good health except for a persistent 50% left-sided perceptive hearing loss. Cranial nerve function was otherwise intact, and there was no lymphadenopathy or hepatosplenomegaly.

Discussion

Sinus histiocytosis with massive lymphadenopathy was first described by Rosai and Dorfman in 1969. Since the initial series of four patients, more than 150 cases have been collected. The disease is found throughout the world. It occurs most commonly in black children; 8% of patients are below 20 years of age. The male to female ratio is 2:1. Massive, symmetrical, painless cervical lymphadenopathy is a prominent sign in nearly all cases. Other lymph node groups are frequently involved. Fever and weight loss complete the clinical picture. Laboratory findings include mild anemia, neutrophilic leukocytosis, elevated ESR, and polyclonal hypergammaglobulinemia. The important morphological features are: marked distention of nodal sinuses with mature histiocytes showing phagocytosis of apparently viable lymphocytes, and proliferation of plasma cells.

The natural history of SHML is one of chronicity, with slow resolution over a period of months to several years. Five patients have died, but only one of them as a direct result of his disease. The etiology remains obscure. The clinical picture suggests an infectious process; indeed, elevated Epstein-Barr virus antibody titers have been detected in 15 of 22 cases. Speculation has also revolved around a possible abnormality of the host immune response.

The extra-nodal forms of the disease have attracted increasing attention in recent years. Twenty-eight percent of patients have involvement of one or more extranodal sites, including the orbital soft tissues, eyelid, upper respiratory tract, salivary glands, skin, bone, and testes. Spinal epidural infiltration causing paraparesis has been documented on two occasions. In the first case, reported by Kessler, et al., a 53-year-old man was found to have an epidural tumor extending from C-7 to T-3, and involving vertebral bone. The lesion...
was removed by an emergency laminectomy, with subsequent resolution of the paraplegia. The second report, that of Haas, et al., described an 11-year-old girl with an epidural block at the C-2 and C-5–T2 levels of myelography. Treatment with prednisolone and vinblastine resulted in almost total disappearance of neurological symptomatology, as well as a marked decrease in adenopathy.

Our case presents several noteworthy features. The patient was somewhat older than most cases. The typical clinical and laboratory features of SHML are entirely absent. The disease appears to be confined to the cranial dura, with no involvement of lymph nodes or other extra-nodal sites. Similar instances of extra-nodal disease representing the initial or dominant manifestations of the condition have been noted. This underscores the false assumption inherent in the designation "sinus histiocytosis with massive lymphadenopathy." A nonspecific term such as "benign lymphohagocytic histiocytosis" may be more appropriate.

The other diagnosis considered in this case was that of histiocytosis X. This histioproliferative disorder of unknown etiology comprises three different clinical syndromes: localized eosinophilic granuloma, Hand-Schüller-Christian disease (multifocal eosinophilic granuloma), and Letterer-Siwe disease. Hand-Schüller-Christian disease, in its classical expression, produces multiple lytic skull lesions which may involve the central nervous system by compression or invasion of contiguous structures. Less frequently the dura itself is the site of the deposits, which may attain a considerable size. Interestingly, the dura of the middle cranial fossa is the most frequent locus of involvement.

Primary growths within the substance of the brain or spinal cord are rare, and are thought to arise from the adventitial cells of blood vessels. In this regard, the well recognized predilection of this disorder for the hypothalamic-pituitary axis has been attributed to the rich vascularity of this region. Histologically, the infiltrate in histiocytosis X contains significant numbers of eosinophils. Lymphohagocytosis is absent. Ultrastructurally, Langerhans' granules are identified. The lesion in our case exhibited none of these features.

With few exceptions, medical therapy for SHML has proven fruitless. It would appear that treatment, if required, should be limited to measures designed to preserve the functional integrity of the affected part. In our case, surgical excision produced symptomatic improvement. The extra-nodal form of SHML has now taken its place in the long list of intracranial space-occupying lesions. As clinicians and pathologists become more familiar with this peculiar disease and its varied manifestations, it is expected that additional cases with neurological involvement will come to light.

Acknowledgment

The author acknowledges Dr. Ronald F. Dorfman, who reviewed the pathological material and confirmed the diagnosis.

References


Manuscript received January 27, 1983. Accepted in final form October 28, 1983.

Address reprint requests to: Michel Trudel, M.D., F.R.C.P.(C), Department of Pathology, McGill University, 3775 University Street, Montreal, Quebec H3A 2B4, Canada.