Systemic Weber-Christian disease presenting as an intracranial mass lesion

Case report

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A case is reported in which systemic Weber-Christian disease presented as a dural mass causing the signs and symptoms of increased intracranial pressure. The literature and possible pathogenesis of this entity are discussed.

KEY WORDS • panniculitis • Weber-Christian disease • lipogranuloma • tumor • dura

REAPING nodular, febrile, nonsuppurative panniculitis was first described in 1892 by Pfeifer,18 and a second case was presented by Gilchrist and Ketron8 25 years later. Weber4 called it "relapsing nodular, nonsuppurative panniculitis," and Christian7 stressed the febrile nature of the disease. In 1936, Brill4 coined the eponym "Weber-Christian disease."

This disorder probably represents a group of diseases of unknown etiology that present in a number of forms: 1) A cutaneous form that most often occurs in women, characterized by recurrent episodes of fever, malaise, and arthralgias, associated with small, usually tender, subcutaneous nodules in the thighs, buttocks, and abdominal wall. 2) A visceral form14 involving the fatty tissues of the gastrointestinal tract and occasionally the retroperitoneal structures. It may present without dermal involvement. 3) A more diffuse "systemic" type19 that may include the organs of the thoracic cavity and bone marrow.

The present patient represents the second case of Weber-Christian disease with intracranial dural involvement, and the first case to have presented as an expanding mass large enough to cause increased intracranial pressure. Arnold and Bainborough5 presented the first case with intracranial dural involve-

FIG. 1. Computerized tomography scan, with contrast enhancement, demonstrating the multilobulated mass.
Intracranial tumor in Weber-Christian disease

Case Report

This 47-year-old man was in good health until 2 months before admission, when he developed progressive dysarthria, left-sided incoordination and impairment of gait. During the same period he also developed diplopia, paralysis of upward gaze, bilateral proptosis, and hypertension.

Examination. Laboratory studies disclosed hypoalbuminemia, hypogammaglobulinemia, a subnormal IgA, and an elevated cerebrospinal fluid protein (104 mg %). A computerized tomography (CT) scan (Fig. 1) disclosed a left cerebellar and quadrigeminal plate lesion which enhanced with contrast material, but was avascular angiographically. Further workup demonstrated cardiomegaly, interstitial pulmonary fibrosis, hydronephrosis, and hepatosplenomegaly, as well as lytic lesions in the left humerus and both femoral heads. Biopsies of the retro-orbital fat, maxillary sinus, subcutaneous nodules of the thigh, and a penile ulcer revealed non-specific inflammatory changes and fibrosis.

The patient was given a trial of Cytoxan (cyclophosphamide) and steroids, and received radiation therapy to the posterior fossa. After initial improvement, he experienced an exacerbation of the dysarthria and hemiparesis, and later became obtunded. A follow-up CT scan disclosed marked enlargement of the lesion with associated hydrocephalus.

Operation. A ventriculopleural shunt was placed, and a left suboccipital craniectomy was performed with subtotal removal of the lesion. The tumor was firm, avascular, encapsulated and yellowish on section. It appeared to arise from the tentorium and extended through the incisural notch.

Postoperative Course. After transient minor improvement, the patient continued on a downhill course marked by progressive obtundation and cardiac arrhythmias. He died 8 days after surgery, 6 months after the onset of his first neurological symptoms.

Postmortem Examination. There was disseminated Weber-Christian disease affecting multiple organs, including those of the retroperitoneum, heart, pericardium, and lungs. Examination of the brain disclosed a large, firm, yellowish lobulated dural mass extending above and below the tentorium (Fig. 2). The adjacent dura and venous sinuses were diffusely infiltrated by the same yellow fibrous fatty tissue that was noted in the viscera, causing almost complete luminal obstruction at the torcula and sinus rectus. The brain parenchyma and cortical vessels were normal.

Microscopically (Fig. 3) there were areas of amorphous eosinophilic material markedly positive to fat stains and the Schultz reaction for cholesterol esters. Clusters of pleomorphic and often bizarre-shaped cells, polymorphonuclear giant cells, and occasional plasma-cell and lymphocytic infiltrates were

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FIG. 2. Autopsy specimen with the tumor left in situ after removal of normal brain. The tumor infiltration of the tentorium can be seen (large arrows). Open arrow indicates the posterior falx, and the curved arrows show the supra- and infratentorial portions of the tumor.

FIG. 3. Photomicrograph of the tumor. A prominent multinucleated giant cell can be seen in the upper left corner. H & E, × 70.
The occurrence of Weber-Christian disease within the cranial cavity is rare. This incidence is thought to be related to the paucity of intracranial lipomatous tissue, which is normally present only as a constituent of the pia mater. Xanthomatous tumors of the dura and meninges have been noted, and a case of a dural hibernoma has been reported. According to Kleinsasser, lipomas per se of the dura have not been described. Schoen, et al., found a cerebellopontine angle "lipoma" in a case of Weber-Christian disease, and Arnold and Bainborough reported a dural "lipoma" with lipogranulomatous changes in another.

The diagnosis of Weber-Christian disease is made by light microscopic findings characteristic of lipogranulomatosis. These findings include areas of fat necrosis surrounded by granulation tissue with numerous mononuclear inflammatory cells, lipophages, and multinucleated giant cells. Biopsy specimens may show the nonspecific changes of chronic inflammation and fibrosis, which probably represent a late or resolution stage of the disease.

There is no satisfactory treatment for the disease, although various regimens including antibiotics, chloroquine, steroids, cyclophosphamide, and radiation therapy have been suggested. The dural mass in the present case grew dramatically in size over a short period of time in spite of trials of steroids, cyclophosphamide, and radiation therapy. Neither the etiology nor the pathogenesis of this entity have been satisfactorily elucidated. Previous studies have failed to demonstrate an infectious or toxic etiology, and it is possible that the disease might be the result of an enzymatic change, or is part of some autoimmune reaction.

The pathogenesis of such a fatty mass in the dura is open to several possibilities. 1) It might arise from a pre-existing lipoma. Arnold and Bainborough stated that the dural tumor they reported must have been a lipoma that had undergone necrosis, just as the fat in other body cavities had done. 2) The mass might originate from normal fat cells in the pia mater which proliferate and extend through the subarachnoid space and into the dura; however, no subpial involvement was noted in this case. 3) Milner and Mitchinson suggested that this entity might result from fat emboli with mesenchymal reaction; however, such an explanation is unlikely as the disease was limited to the dura. 4) The mass might have developed in continuity with the diffusely involved dural sinuses. 5) It could have arisen from relatively undifferentiated mesenchymal cells, which might be stimulated or be genetically predestined to undergo proliferation along lipomatous lines. This latter hypothesis seems most tenable, as the staining characteristics of the fatty mass included those of cholesterol esters, implying xanthomatous change rather than lipomatous degeneration.

Whether the dural tumor found in this case represents an incidental finding (such as a dural lipoxanthoma) associated with Weber-Christian disease, or is simply part of the generalized disease process is not clear. Also, the possibility of a relationship between this entity and the reticuloendothelioses, such as Hand-Schüller-Christian disease and Letterer-Siwe disease, which have similar clinicopathological features, leaves the status of the "systemic" form of Weber-Christian disease an open question.

Discussion

References

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