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CT of CNS Lesions in Lymphomatoid Granulomatosis: Case Report

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Neurologic manifestations of lymphomatoid granulomatosis occur in 20–30% of cases. The disease is primarily an unusual form of pulmonary angitis, and was first described in 1972 by Liebow et al. [1]. The purpose of this paper is to show the progression and regression of CNS lesions in a 17-year-old boy who presented only with neurologic features of the disease.

Patients with lymphomatoid granulomatosis are usually quite ill with pulmonary manifestations. In our study, lymphomatoid granulomatosis initially manifested purely as a CNS disease that was later followed by histologically proved abdominal involvement and radiologic evidence of chest involvement.

Case Report

A 17-year-old boy presented with signs and symptoms of a left-sided posterior fossa neoplasm in October 1981. CT revealed a large, left-sided, contrast-enhancing neoplasm surrounded by mild edema. The neoplasm was somewhat diffuse in appearance and had no sharply defined margins. Slight compression of the fourth ventricle was noted and this was pushed across to the right side (Fig. 1). The provisional scan diagnosis was that of an astrocytoma. However, other posterior fossa neoplasms were also included in the differential diagnosis. A suboccipital craniotomy was performed and the neo-

plasm was partially removed. No radiotherapy or steroids were administered at this stage. The patient had an uneventful recovery after surgery, and follow-up CT scans 4 weeks after presentation showed a residual area of contrast enhancement at the site of surgery (Fig. 2).

Follow-up CT scans 2 and 21 months after surgery showed marked regression of the original lesion, which had totally resolved 21 months after presentation.

Thirty-five months after presentation, the patient presented with symptoms and signs of a right-sided cerebellar neoplasm. A scan at this time was similar in appearance to the first scan, although more diffuse in nature (Fig. 3). No other intracranial lesions were present. Through a suboccipital approach, the lesion was resected. After a course of radiotherapy, the patient made an uneventful recovery.

Thirty-nine months after presentation, the patient experienced slurring of speech and severe headaches. A CT scan 2 months later revealed no evidence of the previous posterior fossa lesions. However, three small supratentorial enhancing lesions were noted adjacent to the body of the right lateral ventricle. Their appearances suggested secondary deposits, and they were surrounded by prominent edema (Fig. 4). These lesions persisted and were surgically removed 42 months after presentation. The patient progressed favorably.

Forty-seven months after presentation, an enlarged spleen was noted. This was followed by a laparotomy, during which a splenectomy, liver biopsy, and resection of mesenteric lymph nodes were performed. The spleen was enlarged with a mass of 643 g, and histologic examination showed involvement of the spleen and the liver by lymphomatoid granulomatosis. The patient remained asymp-

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Fig. 1.—Initial CT scan with contrast. Left cerebellar lesion shows no clear-cut margin.

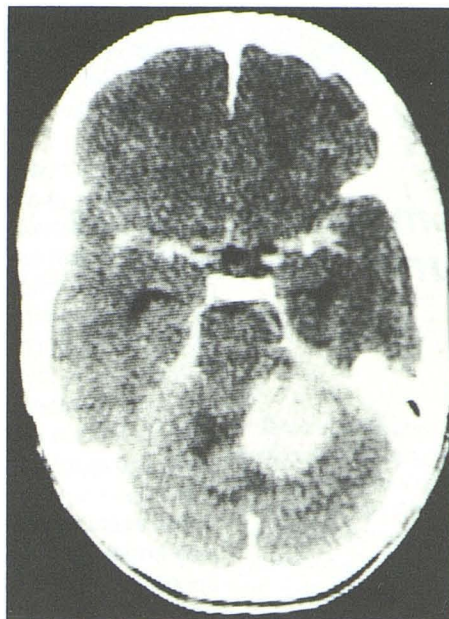


Fig. 2.—Postcraniotomy follow-up CT scan of left cerebellar lesion shows poorly defined contrast enhancement.



Fig. 3.—Follow-up scan 35 months after presentation shows right cerebellar lesion.

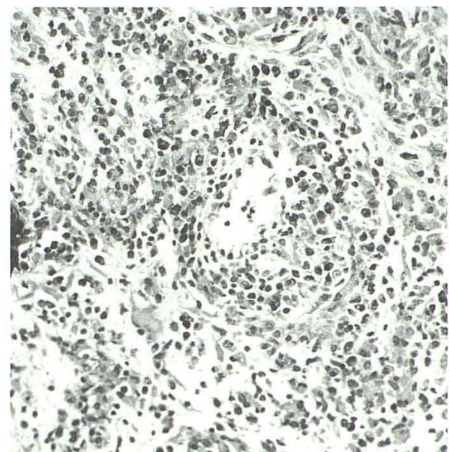
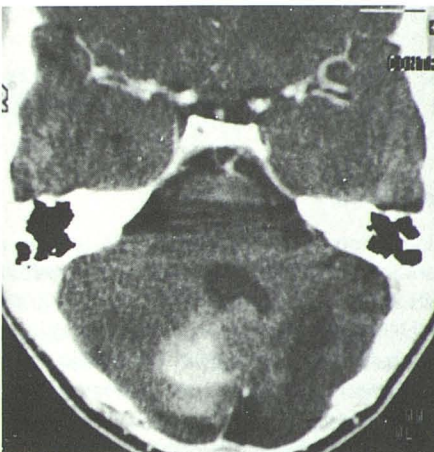


Fig. 4.—CT scan shows three small supratentorial infiltrates.



tomatic and treatment has been maintained on corticosteroids and cytotoxic drugs.

CT scans performed 48 months after presentation showed the cerebral lesions to have disappeared completely. Scanning of the abdomen revealed no lymphadenopathy, but the lung scan showed a small lesion in the apex of the right lung with slight infiltration, although the patient was asymptomatic.

The most recent scans of the brain and chest, 51 months after presentation, showed no active or residual lesions.

The CT appearances mimicked those of primary cerebral neoplasms, although metastatic carcinomas and lymphoma are included in the differential diagnoses. Although the CT features were not pathognomonic, the progression, regression, and appearance of multiple CNS lesions strongly favored lymphomatoid granulomatosis.

Results

The biopsy from the left cerebellar lesion consisted of white matter and a segment of granule cell layer. Within the white

matter there was a diffuse and moderately intense proliferation of astrocytes with abundant pink cytoplasm, which showed little nuclear pleomorphism. A polymorphic infiltration around blood vessels was present, consisting of lymphocytes, plasma cells, and histiocytes. The cells were seen to invade the vascular walls, but no necrosis or granuloma formation was apparent (Fig. 5). No atypical lymphoreticular cells were present.

The histopathologic features of the right cerebellar lesion were similar to those of the original lesion in the left cerebellar hemisphere, with the exception that the mononuclear infiltration, and especially the plasma cells, were more prominent. The features in the right cerebral biopsy were similar to those noted in the cerebellar biopsies.

The analogous histologic character of the lesions in the cerebellum and in the cerebrum suggested a single disease process, consistent with that of lymphomatoid granulomato-

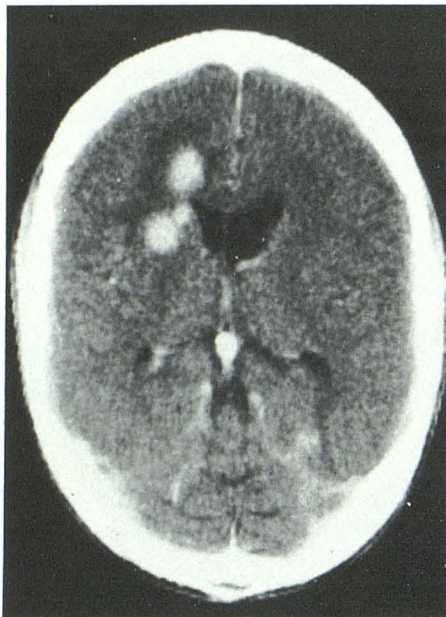


Fig. 5.—Polymorphic perivascular infiltration and astrogliosis (H and E $\times 313$).

sis. Immunohistochemistry showed the cellular infiltration to be polyclonal and therefore consistent with an inflammatory disease.

Discussion

Lymphomatoid granulomatosis usually presents as a primary lung affliction, with secondary metastatic spread to the CNS. Initially, however, lymphomatoid granulomatosis can be clinically confined to the CNS, although rarely [2–5]. The diagnosis can be made in the absence of usual pulmonary involvement, and cerebral involvement may occur up to 5 years before detection of pulmonary disease [6]. In our patient with histopathologically proved cerebral lymphomatoid granulomatosis, there was no evidence of extracranial involvement until 4 years later, when splenomegaly was found, and in the fifth year, when an isolated solitary lung lesion was present.

Treatment of lymphomatoid granulomatosis in the CNS has been unrewarding. Lymphomatoid granulomatosis generally has a poor prognosis, with a mortality rate of 60–90%, and neurologic manifestations are considered a grave prognostic sign [6]. Treatment has centered on the use of steroids and cytotoxic agents, but the response of CNS lesions to these substances has been disappointing [6]. Cranial irradiation has been used for focal lesions, and the results have been variable, with some patients responding to such treatment [7, 8]. However, a favorable outcome after surgical treatment alone has been reported [4].

In contrast to the poor results previously documented, our patient seems to have responded well to intracranial surgical treatment. The outcome to date suggests that the prognosis for lymphomatoid granulomatosis, when confined to the brain initially, is considerably better than when it is part of the generalized disease. CT scan appearances should be differ-

entiated from (a) primary and secondary brain neoplasms, (b) multiple secondary metastatic deposits, and (c) lymphomatous or sarcomatous infiltrates. Although the lesion can be adequately demonstrated on CT, the final diagnosis remains histologic.

Immunohistochemistry for monoclonal light chains and heavy chains showed the cellular infiltrate to be polyclonal, indicating a nonclonal proliferation of plasma cells, which is consistent with an inflammatory rather than a neoplastic process [9]. Lymphomatous transformation can occur in up to 20% of patients [10], and this is heralded by monoclonal immunoglobulin production in the cellular infiltrate [9].

Prognosis is favorably influenced by the histologic criterion of a large number of small lymphocytes and histiocytes. An adverse feature is a large number of atypical lymphoreticular cells. The extent of necrosis did not influence the prognosis [6].

Our case emphasizes that lymphomatoid granulomatosis may present as single or multiple intracranial lesions, the CT appearance of which may be easily confused or misinterpreted for other primary or secondary neoplastic diseases. This case history underlines the idea that lymphomatoid granulomatosis rarely involves primarily the CNS. For example, associated lesions, such as splenomegaly and pulmonary lesions, as in our patient, are almost always present although they may be clinically undetectable or develop later during the course of the disease. Finally, the history of our patient confirms that it is possible to clear the intracranial lesion by excision, followed by radiotherapy and steroid administration.

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