The numerous eponyms and terms that have resulted from the study of sarcoidosis over the years reflect the protean clinical manifestations of this disease entity. Historical reviews reveal that in 1875 the first clinical report of a case was made by Hutchinson. In 1889 Besnier designated the skin changes "lupus pernio." Boeck (1899) described the histologic features of the cutaneous lesions and introduced the term "sarcoid." Heerfordt (1909) reported the uveoparotid fever syndrome, which later became identified as a manifestation of sarcoidosis. Schaumann (1914) was first to recognize the systemic nature of the disorder and suggested the term "benign lymphogranuloma." Osseous lesions, previously recognized by other observers, were established as part of the generalized disease by Jüngling in 1919.

Though extensive investigations as yet have failed to determine the etiology of sarcoidosis, clinical aspects have advanced impressively, and the cutaneous, visceral and lympho-hematopoietic manifestations have become well established. Neurologic expressions of the disorder, not widely appreciated, are of particular interest. Involvement of the nervous system is relatively uncommon, and intracranial sarcoidosis simulating brain tumor is decidedly rare, only one case being cited by Essellier, et al. in their extensive review of the literature in 1951. Additional cases of sarcoid tumor were recorded by Everts in 1947 and by Höök in 1954.

The present report is concerned with the successful surgical removal of a solitary, circumscribed intracranial tumor resembling a meningioma from the floor of the middle fossa. It was identified histologically as a sarcoid granuloma.

**CASE REPORT**

**History.** A negroid male, aged 19, on active duty in the U.S. Navy, was admitted to the hospital on July 18, 1954, because he had suddenly lost consciousness while aboard ship.

His illness began in July, 1953, when he was bothered by a roaring noise in his right ear. This was continuous and occasionally associated with headache and slight vertigo. In January, 1954, he was hospitalized for treatment of an infection of the right ear, and it was noted that he had polyps in the right external auditory canal. His symptoms continued and he returned to the hospital in April, 1954, when the polyps were surgically removed.

He then began to suffer from almost continuous right-sided headaches with intermittent nausea and vomiting. In addition he had frequent, brief episodes of slight mental confusion associated with unpleasant olfactory hallucinations. Distinct feelings of familiarity occurred at times, even when he was in a strange environment. In the month prior to admission he had two spells, characterized by automatic behavior with subsequent amnesia for his actions. He was found walking aimlessly about the ship, seemingly in a dazed state, and when attempts were made to restrain him he reacted in a maniacal, belligerent manner and finally lost consciousness.

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* The opinions or assertions contained herein are the private ones of the authors and are not to be construed as official or reflecting the views of the Navy Department or the naval service at large.

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Examination. General physical findings were normal. Neurologic examination revealed bilateral papilledema of 4 D. There were small hemorrhages about both discs. Visual fields showed enlarged blind spots. The remainder of the cranial nerves were intact. The motor system showed good power throughout, although on prolonged repetitive movements the left extremities fatigued sooner than the right. There was mild increase in muscle tone in the left extremities. Coordination was normal. Sensation was unimpaired. The deep tendon reflexes were active and equal except that the left quadriceps response was slightly greater than the right. Plantar stimulation produced flexor responses of the toes bilaterally. The left superficial abdominal reflexes were consistently less active than the right.

Routine blood studies and urinalysis gave normal findings. Blood serology was negative. Roentgenogram of the chest showed no abnormalities. Skull roentgenograms revealed an enlarged sella turcica with demineralization of the clinoid processes. Lumbar spinal puncture showed a pressure of 280 mm. of clear CSP. The fluid contained no cells; sugar was 45 mg. per 100 cc., chloride 128 mEq. per liter, globulin 3+, and total protein was 100 mg. per 100 cc.

Electroencephalogram was disturbed by gross focal changes, consisting of higher potentials, slow activity of 1½ to 4 per sec. frequency, irregularity of form and asynchrony which appeared in the anterior temporal, mid-temporal and low frontal areas on the right side (Fig. 1). Right percutaneous carotid arteriography revealed displacement of the middle cerebral artery upward and to the left, indicating the presence of a mass in the right temporal area (Fig. 2).

Operation. Craniotomy was performed on July 26, 1954. The tumor was not immediately evident on the surface of the temporal or posterior frontal lobes but was located by use of the Selverstone-Robinson probe counter, the patient having received intravenous radioactive phosphorus the previous day. Counting in the right temporal lobe delineated an area in which there was a ratio of 70 to 100:1 counts which extended from directly below the middle temporal gyrus medially for a distance of about 5 cm. Removal of a small portion of the inferior right temporal lobe revealed the tumor lying on the floor of the middle fossa. The mass was about the size of a goose egg, measuring 7×5×5 cm., and extended medially almost to the mid-line. Removal of the tissue was accomplished by punch and electrocautery loop. The

![Fig. 1. Electroencephalogram showing high voltage slow activity in the right temporal and low-frontal areas.](image-url)
tumor had a definite capsule, was not invading brain substance, and strongly resembled meningioma. It was fastened very firmly over an area of at least 1 square inch on the floor of the middle fossa. Most of the tissue attached to the floor of the fossa was removed.

*Pathologic Examination. Gross.* The submitted surgical specimens of the right middle fossa tumor consisted of moderately firm pieces of grey tissue. *Microscopic.* Sections of the tumor stained with hematoxylin-eosin showed dense granulomatous tissue composed of numerous small discrete and confluent nodular masses of epithelioid cells separated by lymphoid tissue (Fig. 3a). There were frequent multinucleated giant cells associated with the epithelioid cells. The giant cells had an abundant pink cytoplasm and many small round peripherally arranged nuclei (Fig. 3b). No specific etiologic agents were found. Acid fast stains were negative. There was no evidence of malignancy. Sections of the adjacent brain tissue showed some vacuolization and moderate gliosis as well as marked perivascular lymphocytic infiltration in a few places. The changes in the brain were presumably secondary to the adjacent granulomatous process. *Pathologic diagnosis:* Boeck's sarcoid granuloma.

*Postoperative Course.* Recovery was uneventful except for a mild hemiparesis of the left extremities for 1 month, after which no neurologic abnormalities could be detected. As a result of the histologic diagnosis a number of studies were performed in an attempt to detect any other manifestations of sarcoidosis. Intradermal tuberculin test (1:100) was negative. The Kveim skin test resulted in a negative response. Serial roentgenograms of the chest showed no abnormalities. Roentgenograms of both hands and feet were normal. Sedimentation rate was normal; plasma proteins, 8.2 gm. per cent; albumin, 4.0; globulin, 4.2; serum calcium, 9.0 mg. per 100 cc.; inorganic phosphorus, 4.4 mg. per 100 cc.; alkaline phosphatase, 2.8 Bodansky units. Sections of lymph nodes exised from the left supraclavicular area revealed no abnormalities.

Since discharge in November 1954, the patient has been doing full-time work at an air-station and has remained asymptomatic.
DISCUSSION

The diagnosis of sarcoidosis was not considered preoperatively in this case. At craniotomy the tumor gave the appearance of a meningioma. Subsequent investigation failed to reveal any evidence of systemic disease except for elevation of the serum globulins. Therefore, it cannot be determined conclusively whether the lesion represented an isolated sarcoid reaction or was a localized manifestation of generalized sarcoidosis. An increase in the globulin fraction of the serum proteins, like the negative reaction to intradermal tuberculin, is commonly associated with Boeck's sarcoid but is not a pathognomonic finding. In regard to the Kveim skin test, various investigators have reported on the incidence of negative reactions in patients with proved sarcoidosis, with figures ranging from 8 to 35 per cent.

The technic of employing radioactive phosphorus and a needle probe counter for brain tumor localization has been established as a distinct diagnostic aid and in this case was of considerable value. The ratio between the count obtained in the area of the tumor and that obtained in the uninvolved brain varied from 70:1 to 100:1.

It is believed that the mass may have been enveloped by thinned dura mater.

Fig. 3. Photomicrographs of tumor. (a) Dense granulomatous tissue composed of numerous small nodular masses of epithelioid cells separated by lymphoid tissue. Hematoxylin-eosin stain, ×155. (b) Single Langhans giant cell; no evidence of necrosis, ×537.
rather than by a true capsule, such as that which was described in the case report of Coleman and Meredith.

Many recorded cases of sarcoidosis have lacked neurohistologic verification, the nature of the nervous system lesions being assumed on the basis of proved sarcoid elsewhere in the body.\textsuperscript{17,18,25,36} Paresis of the facial muscles is the commonest sign of peripheral nerve implication, although any of the cranial nerves may be affected,\textsuperscript{9} and there may be widespread neural involvement. The clinical expression of intracranial sarcoidosis is variable and depends upon the site of the infiltration. Diabetes insipidus has been a prominent symptom in about one-third of the recorded cases,\textsuperscript{25} while convulsions, visual field defects, papilledema, spastic hemiplegia, hemianesthesia, cerebellar signs, mental aberrations, disturbances of sleep, headache, aphasia, and a variety of other cerebral signs and symptoms also have been described.\textsuperscript{18,19,25,28,36}

Most of the pathologically established cases of central nervous system sarcoid have shown variable degrees of meningoencephalomyelitic infiltration, particularly of the basal structures. Cases of autopsy-proved meningoencephalitis caused by sarcoidosis have been reported by Erickson, Odom, and Stern,\textsuperscript{6} Essellier, Koszewski, Lüthy, and Zollinger,\textsuperscript{7} and Ricker and Clark.\textsuperscript{57} Colover\textsuperscript{4} recorded a case of posterior fossa arachnoiditis with fourth ventricle and cerebellar adhesions associated with sarcoid infiltration. Naumann\textsuperscript{24} described sarcoid pachypleptomeningitis in an infant. Hypophysial infiltration characterized the case reported by Tillgren,\textsuperscript{25} while the case of Gjersøe and Kjerulf-Jensen\textsuperscript{12} showed involvement of the hypothalamus but not of the pituitary. Coleman and Meredith\textsuperscript{8} recorded a case in which an encapsulated pituitary mass was surgically removed; it was initially reported as diffuse tuberculosis of the pituitary gland, but subsequent histologic study established the diagnosis of Boeck’s sarcoid. Aszkanazy\textsuperscript{1} recorded 5 cases: 1 with meningo-myelitis of the upper thoracic cord; 1 with meningoencephalitis of the right pre- and postcentral gyri; 1 with pituitary invasion; and 2 with sarcoid tissue extending from the optic chiasm into the floor of the third ventricle. In 3 of these cases the disease could not be demonstrated elsewhere in the body. A case presented by Lenartowicz and Rothfeld\textsuperscript{19} in 1930 and by Reis and Rothfeld\textsuperscript{26} in 1931 revealed extensive sarcoid involvement of the cerebellum, infundibulum, floor of the third ventricle, both caudate nuclei, optic chiasm and nerves, anterior commissure, right olfactory bulb and gyrus rectus, left lenticular nucleus and left temporal lobe.

There have been very few reports of isolated tumors. Höök\textsuperscript{18} described one patient with systemic sarcoidosis in whom a small tumor arising in the choroid plexus filled the upper portion of the inferior horn of the right lateral ventricle. It was extirpated and grossly resembled a choroidal papilloma; microscopic study established the diagnosis of sarcoid. Another case of generalized sarcoidosis presented by Höök lacked neurohistologic verification but there was clinical evidence of an expanding mass of the left frontal lobe and Sylvian fissure which disappeared under conservative medical management; such spontaneous and often dramatic remission of signs and symptoms is a characteristic feature of sarcoidosis. A patient of Everts\textsuperscript{8} had a large left occipital lobe tumor which was successfully removed and histologically proved to be sarcoidosis; similar lesions were found in the posterior nasal pharynx, and roentgenograms of the chest revealed a left hilar mass. The case of Uehlinger (cited by Colover\textsuperscript{1} and by Essellier et al.\textsuperscript{7}) clinically demonstrated a mid-
brain syndrome; at autopsy there was a circumscribed mass at the base of the brain which histologically was sarcoid granuloma.

Emphasis has been placed on the cerebrospinal fluid changes in sarcoidosis. Pleocytosis and an increase in protein content should arouse suspicion of sarcoid of the central nervous system in cases in which there is evidence of the disease elsewhere. A decrease in sugar content has also been reported. The spinal fluid in the present case contained no cells, elevated protein and lowered sugar content.

**SUMMARY AND CONCLUSIONS**

The case reported here clinically represented an expanding lesion of the right temporal lobe as manifested by olfactory hallucinations, psychomotor seizures, déjá vu phenomena, papilledema and slight left-sided signs of pyramidal tract dysfunction. Skull roentgenograms revealed enlargement of the sella turcica and demineralization of the clinoid processes. Electroencephalogram showed changes compatible with a gross irritative focus in the right temporal area. Elevation of the right middle cerebral artery, as demonstrated by carotid arteriograms, was indicative of a mass in the temporal lobe. At operation a large tumor with the gross appearance of a meningioma was found projecting upward from the floor of the middle fossa. It compressed the inferior portion of the temporal lobe, appeared to be encapsulated, and did not invade the substance of the brain. It was completely removed and histologically proved to be sarcoid granuloma. Subsequent investigation directed toward identification of systemic disease was negative except for elevation of the serum globulins.

Sarcoidosis of the central nervous system, though relatively uncommon, should be considered as a diagnostic possibility in patients with neurologic abnormality, including signs of cerebral tumor, who show evidence of sarcoid elsewhere in the body. Rarely, an intracranial lesion may represent the only discernible expression of the disease process.

**REFERENCES**


