Sarcoidosis of the pineal gland

Case report

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The authors report a case of sarcoidosis of the pineal gland with no other organ manifestation of the disease. The tumor was successfully removed by a direct surgical approach.

KEY WORDS • pineal gland • sarcoidosis • obstructive hydrocephalus • computerized tomography • right occipital approach

In recent years teratomas, pinealomas, gliomas, and cysts of the pineal region have to an increasing extent been operated on by the direct approach.\(^1\)\(^-\)\(^7\)\(^,\)\(^9\)\(^,\)\(^10\) In a survey of the literature we have found no case of sarcoidosis (Besnier-Schaumann-Boeck disease) located in and confined to the pineal body.\(^8\)

Case Report

This 13-year-old boy was referred to us on June 11, 1975, because of suspected obstructive hydrocephalus. There was a history of intermittent headaches since January, 1975, and of recent disturbance of vision.

Examination. On examination the following abnormal findings were noted: marked bilateral papilledema; bilateral oscillating horizontal nystagmus; and a positive Babinski reflex on the right side. The electroencephalogram was generally altered without focal findings. Blood chemistry and x-ray studies of the lungs were unremarkable. Computerized tomography (CT) with intravenous injection of contrast medium (Telebrix) showed a median tumor, 2.5 cm in diameter, in the pineal region, and obstructive hydrocephalus. Ventriculography with positive contrast medium (Dimer X) confirmed this finding. In addition, very fine calcifications were found in the tumor.

On June 12, 1975, a left ventriculoatrial shunt with a Holter valve was inserted. The symptoms of increased intracranial pressure rapidly regressed. One month later, papilledema could no longer be demonstrated.

Operation. In a second session on September 2, 1975, one of us (C.L.) operated on the tumor directly. A right occipital approach was selected with the patient in a sitting position. Anesthesia was performed using neuroleptic analgesia and positive end-expiratory pressure (PEEP) respiration. The electro-
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cardiogram, central venous pressure, and arterial pressure were continuously monitored. After mobilizing and lifting the right occipital lobe, the tentorium was incised longitudinally and parallel to the sinus rectus. The blood vessels running from the cerebellum to the caudal tumor pole were bipolarly coagulated. After incision of its capsule, the plum-sized sparsely vascularized tumor could be completely removed without disturbing the ampulla Galeni.

**Histological Finding.** The histological finding was sarcoidosis (Fig. 1). The tissue was almost exclusively granulomatous in nature; remnants of pineal parenchyma could be discerned in a marginal region. Large areas were intensely sclerosed. The granulomas were composed of aggregates of epithelioid cells and histiocytes with lymphocytes and connective tissue cells at the periphery, often forming reactive follicles. Many of them showed large multinucleated giant cells with characteristic cytoplasmic inclusions of two types: either light-yellow, highly refractile, amorphous material, or hematoxyphilic calcifications with concentric or polycentric lamellar layers. Such Schaumann bodies sometimes also lay isolated in the tissue.

**Postoperative Course.** Postoperative neurological symptoms were confined to a transient vertical visual paresis and a transient psycho-organic syndrome. On September 22, 1975, the boy was discharged in good condition. In December, 1975, he returned to school and has been attending since, without neurological difficulties.

One year after operation, the neurological findings are within normal limits. Bodily growth and secondary sex characteristics are normally developed according to age. There is no indication for any other organ manifestation of sarcoidosis. A control CT scan shows a ventricular system of normal width and no indication of recurrent tumor.

**Discussion**

Space-occupying lesions in the pineal region are very rare. According to Zülch, they only amount to 1.2% of all brain tumors. In our case the symptoms were restricted to those of increased intracranial pressure. The CT scan, by precisely giving the midline location and size of the tumor, as well as the ventricular width, was of crucial significance in arriving at the diagnosis. Ventriculography did not yield any additional information in our case. The strictly median position and the absence of signs of lateral infiltration into the thalamus are cogent criteria for surgery.

The histological diagnosis was a surprise since the patient showed no other organ manifestations of sarcoidosis. We can, however, report one comparable case from our department. This patient was a young woman with sarcoid granulomas in the right temporal lobe. After operation in that case, there has also been no recurrence of the intracranial tumor nor any other organ manifestation of sarcoidosis. The prognosis of these isolated central nervous system granulomas appears favorable when totally removed. We feel that a direct surgical approach is justified even in cases of difficult access like the pineal region.

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Fig. 1. Section of biopsy from pineal tumor, showing granulomatous tissue with scattered giant cells. H & E, × 100. Inset: Multinucleated giant cell with strongly double refractile inclusions. Polarized light, × 250.
References


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