Haemangiopericytoma of the pineal body

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Less than 1% of neoplasms of the central nervous system arise in the pineal body (Kernohan and Sayre, 1952). The great majority of these are germ cell neoplasms, germinomas (seminomas), and teratomas (Russell, 1944; Friedman, 1947; McGovern, 1949; Dayan, Marshall, Miller, Pick, and Rankin, 1966). Gliomas, true pineocytomas, and meningiomas are much less common (Russell and Rubinstein, 1963). Single examples of ganglioneuroma (Horrax and Bailey, 1928) and chemodectoma (Smith, Hughes, and Ermocilla, 1966) have been noted. Haemangiopericytomas have been reported in the meninges of brain and spinal cord and within the brain (Fisher, Davis, and Lemmen, 1958; Kruse, 1961; Kernohan and Uihlein, 1962; Pitlyk, Dockery, and Miller, 1965) but we are unaware of any previously reported example arising in the pineal body.

CASE REPORT

A 28-year-old right-handed janitor was admitted to The University of Michigan Medical Center complaining of having had severe frontal headaches for one month. He had been well until four months previously, when he noted loss of energy and the family noted personality changes. During the three weeks before admission he vomited occasionally and developed intermittent double vision, decreasing vision in the left eye, and increasing mental confusion. He had loss of recent memory and difficulty in recalling details of his present illness.

Physical examination showed a well-developed, muscular white male whose speech was occasionally slurred. The pupils were dilated and reacted well to light and accommodation. Horizontal nystagmus was noted on the left. There was bilateral papilloedema, more severe on the left. No functional deficit of the remaining cranial nerves was noted. A Babinski sign was present on the right but was questionable on the left. The right ankle and knee jerks were increased and there was terminal tremor on finger to nose test which was worse on the right. Strength in all extremities was normal and there was no sensory loss. The remainder of the physical examination was normal.

The haematocrit was 49% and the white blood cell count, prothrombin concentration, blood urea nitrogen, and two-hour post-prandial blood sugar were normal, as were radiographs of the skull and chest. A brain scan using 700 μ c mercury²⁰³ showed a midline lesion approxi-

mately 6 × 6 × 4.5 cm (Figs. 1 and 2). A ventriculogram showed a soft tissue mass 5 cm in diameter in the region of the pineal body. Cerebrospinal fluid (CSF) taken by ventriculostomy contained gross blood. The Kahn test on the CSF was non-reactive, the total protein 80 mg/100 ml. and the colloidal gold reaction 1123210000.

Immediately after the ventriculogram a right posterior craniotomy was done. A pink neoplasm, 5 cm in diameter, was found in the region of the pineal body and the quadrigeminal plate. It was soft, friable, and extremely vascular. The pineal body itself was not identified, presumably due to replacement and destruction by the neoplasm which was totally removed in pieces with considerable accompanying loss of blood. Post-operatively the patient breathed spontaneously but had intermittent Cheyne-Stokes respirations. He never regained consciousness and died 12 hours after completion of the operation.

PATHOLOGICAL FINDINGS Histological sections of formalin fixed tissues were stained with haematoxylin and eosin, phosphotungstic acid haematoxylin and by Cajal's gold sublimate method and Wilder's silver impregnation method for reticulin. Frozen sections of fresh tissue were stained by the silver carbonate method of del Rio Hortega.

FIG. 1. Brain scan in the left lateral position using 700 μ c mercury²⁰³. A 4.5 cm × 4.5 cm area of increased uptake is present near the midline.

FIG. 2. Brain scan in the posterior position shows a 6 cm $\times$ 6 cm midline area of increased uptake of the radioactive material.

The neoplasm consisted of clusters of relatively uniform oval and polygonal cells with round and oval nuclei, and small to moderate amounts of cytoplasm. There was little cellular and nuclear pleomorphism. The nucleoli were small and the chromatin granular and well dispersed. Normal appearing division figures were scattered uniformly throughout, averaging one for every two high power fields ($\times 450$) (Fig. 3). Scattered nuclei were shrunken, hyperchromatic, and irregular. A rich network of dilated and collapsed sinusoidal spaces lined by single layers of flattened endothelial cells separated the neoplastic cells into clusters and festoons, producing an organoid pattern in some areas (Figs. 4 and 5). Reticulin stains confirmed the peritheliomatous position of the neoplastic cells to blood vascular spaces and showed fine fibres related to many of the individual cells. Del Rio Hortega's silver carbonate, Cajal's gold sublimate, and phosphotungstic acid haematoxylin stains did not reveal any bulbous processes or nerve fibrils, which are a feature of normal cells in the pineal body. The histological characteristics were those of a haemangiopericytoma.

A necropsy done four hours after death revealed extensive haemorrhage into the lateral, third, and fourth ventricles. No residual neoplasm was found but there was

FIG. 3. Neoplasm with a single mitotic figure just below the centre of the field. Haematoxylin and eosin. $\times 500$.

FIG. 4. Neoplasm with a prominent sinusoidal pattern adjacent to a more solid appearing area in which the sinusoids are collapsed. Haematoxylin and eosin. $\times 120$.

a mass of clotted blood 7 cm in greatest diameter in the area of resection.

DISCUSSION

Stout and Murray first defined the haemangiopericytoma as a definite entity in 1942 and many cases have since been reported in a variety of locations. Most of these have been located in the

soft tissues, but since 1949 (Stout) a small number of cases have been reported in the central nervous system. We found reports of 45 such tumours, specifically designated as haemangiopericytoma, in the English literature: 40 were intracranial and five intraspinal. Most neoplasms have had attachments to the meninges but a few have been within the substance of the brain (Peace, 1954; Kernohan and Uihlein, 1962) or in the lateral ventricle

FIG. 5. Neoplasm in the field as Figure 4. A single layer of endothelial cells separates the pericytic cells from the vascular lumina. This pattern is present in areas of sinusoidal collapse as well as where there is sinusoidal dilatation. Haematoxylin and eosin. $\times 220$.

(McDonald and Terry, 1961) with no apparent meningeal connection. The commonest locations have been the frontal or parietal regions of the cerebrum but at least nine were in the posterior fossa and several were temporal or occipital in location and one was on the floor of the middle fossa (Solitare and Krigman, 1964). None have been reported previously in the region of the pineal body.

The existence of haemangiopericytoma in the central nervous system has been debated. Begg and Garret (1954) considered haemangiopericytomas to be identical histologically with the angioblastic meningiomas reported by Bailey, Cushing, and Eisenhardt in 1928 and again by Cushing and Eisenhardt in 1938. Russell and Rubinstein (1963) concluded that there were no convincing grounds for separating haemangiopericytoma from meningiomas in general. The considerably poorer prognosis for patients with so-called angioblastic meningiomas as compared with classical meningiomas as well as their different histological appearance would appear to justify their separation as distinctly different neoplasms, and when angioblastic meningiomas are compared with haemangiopericytomas of soft tissues they would appear to be the same. Ramsey (1966) described the ultrastructure of a haemangiopericytoma of the central nervous system and contrasted it with that of a haemangioendothelioma of brain. Similar studies have not been made on a so-called angioblastic meningioma. Kernohan and Uihlein (1962) regarded haemangiopericytoma of the central nervous system as a definite pathological entity separate from meningotheial meningioma, although they reserved judgement as to the cell of origin.

Haemangiopericytomas are presumed to arise from the pericytes of Zimmerman (Stout and Murray, 1942). Pericytes have been identified in the pineal body of rats (Gusek and Santoro, 1961) and cattle (Anderson, 1965) by electron microscopy. Although they have not been specifically demonstrated in the human pineal body, there is no reason to suspect that they are not present.

The neoplasm reported here had many features akin to the chemodectoma of the pineal body reported by Smith, Hughes, and Ermocilla (1966) and, in some fields, it would not be possible to distinguish between the two. However, this lesion lacked the type of organoid and sinusoidal arrangements as well as the cytological features that are characteristic of chemodectomas and paragangliomas.

The interpretation of benignancy or malignancy on the basis of histological criteria is difficult in many haemangiopericytomas. This pineal neoplasm was clinically malignant by virtue of location and fairly

rapid clinical course, but whether it would have recurred or metastasized had the patient survived is unanswerable. However, because of the scattered division figures, the degree of cellularity and some nuclear pleomorphism, it probably should be regarded as at least potentially malignant.

SUMMARY

An unusual vascular neoplasm which arose in the region of the pineal body of a 28-year-old man is described and interpreted as a haemangiopericytoma. Haemangiopericytomas of the central nervous system are uncommon and this is the first report of such a tumour in the pineal body. Patients with intracranial haemangiopericytomas have a considerably poorer prognosis than those with meningiomas.

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