Ectopic Pinealoma: Report of Three Cases.

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Citation: 日本外科宝函, 1953, 22(2): 145-147

Issue Date: 1953-03-01

URL: http://hdl.handle.net/2433/205977

Type: Departmental Bulletin Paper

Textversion: publisher

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(Received for publication, Jan. 9, 1953)

Case Report

Case 1, A. T., a man aged 28. Subcortical cyst with a mural nodule of typical pinealoma in the left frontal lobe. Tumor of the pineal body was excluded by the iodized oil ventriculography.

The patient was admitted to our hospital complaining of headache, right hemiplegia and of difficulty in speech. Tumor in the left cerebral hemisphere, maybe in the frontal lobe, was suspected from neurologic findings and was verified by the iodized oil ventriculography.

Left frontal craniotomy was performed. By exploratory puncture through the dural membrane, clear fluid of straw-yellow color was obtained at a depth of 1 cm. The cortex was incised and the cyst evacuated. It was unilocular with a mural nodule of 3 cm. in diameter at the bottom. The nodule was removed totally.

Microscopic features of the mural nodule: Tumor tissue contains well developed stroma of connective tissue and rather few blood vessels. Tumor cells are characteristic of pinealoma, consisting of admixture of two types of cells, the one being large epitheloid cells and the other lymphoid cells - Fig. 1.

Case 2, K. N., a boy aged 18. Pinealoma or pineoblastoma in the region of the right basal ganglia. No clinical symptoms suggesting a tumor in the pineal region.

He was admitted to the hospital complaining of right hemiplegia of four months duration. It was at first considered, on the basis of clinical findings, there may be a tumor in the right side of the midbrain, but, the correct diagnosis of a tumor in the region of the right basal ganglia was made by the air study.

Right fronto-parieto-temporal flap was turned down. By exploratory puncture, a mass of tumor-like hardness was touched at a depth of 3.5 cm. from the surface. Dissecting the cortex the tumor tissue was curetted subtotally piece by piece. The size of the tumor appeared to be as large as a sparrow's egg.

Microscopic features of the tumor tissue: The tumor is considerably cellular and large cells with round vesicular nuclei are packed together without showing any arrangement or pattern. Some nuclei of these cells contain nucleoli. A few lymphoid cells are scattered between larger cells. The tumor may be a pinealoma or a pineoblastoma - Fig. 2.


She had been suffering from failure of vision and headache since one and a half years before admission. A cerebral tumor was suspected, but the precise localization was obscure.
Definite diagnosis of the left lateral ventricle tumor was obtained by the iodized oil ventriculography.

Left fronto-temporal craniotomy was done. The operation, however, had to be given up without finding out a tumor because of the critical condition of the patient. She died forty three hours after operation.

Autopsy: An oval tumor of $6 \times 4 \times 2.5$ cm. was found in the middle portion of the left lateral ventricle, attaching to its lateral wall. It was grayish red in color and elastic soft in consistency, being covered with thin capsule. The pineal body existed in normal position with normal shape (Fig. 3).

Microscopic findings: The tumor tissue is moderately cellular but not so vascular. Tumor cells are round or oval in shape and almost uniform in size, having scanty protoplasm. Nuclei are vesicular and contain a few chromatin granules. Neuroblasts, spongioblasts and connective tissue can not be found. It can be diagnosed as pineoblastoma (Fig. 4).

Discussion

Above mentioned three cases are all ectopic pinealomas, i.e., case 1 in the left frontal lobe, case 2 in the region of the right basal ganglia and cases 3 in the left lateral ventricle. The presence of a primary pineal tumor can be denied by iodized oil ventriculography in case 1, from neurologic findings in case 2, and by autopsy in case 3.

There may be three possibilities for the occurrence of pinealoma in the unusual, ectopic region.

In the first place a tumor of the pineal body may extend so far to the remote region. NISHINO has reported in detail a case of pinealoma invading extensively the midbrain and the diencephalon.

The second is the metastasis of a primary pineal tumor. In spite of its essentially benign nature, the disseminated metastases of pinealoma in the subarachnoid space or in the ventricle wall by the flow of the cerebrospinal fluid, as in the case of medulloblastoma, are well known. There are considerable number of reports of such cases by BERBLINGER and others.

The third possibility is the primary ectopic occurrence of pinealoma without a tumor of the pineal body itself. AKASAKI, STARK and others have reported such cases. Common location of those ectopic pinealomas is the infundibular region.

Peculiar cases have been described by MACKAY and FORD and MUNCIE in which the whole ventricular wall is replaced by the layer of pinealoma tissue without forming any circumscribed tumor mass. MACKAY states that the pineal body is nothing but a highly differentiated ventricular ependymal tissue and suggests the possibility for arising of pinealoma from any part of the ventricular ependyma. It seems to me that my case 3 offers an evidence for his assumption, because there can be little doubt in the case about the ependymal origin of the tumor, having no contact with the pineal body.

Histogenesis of the ectopic tumor can most easily be attributed to the aberrant embryonic cell rests. Admitting, however, the ependymal origin of pinealoma, as MACKAY suggests, there remains the possibility that in cases 1 and 2, tumors may have arisen from ventricular ependyma and have developed extraventricularly towards the subcortex of the
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As I have previously described, such mode of growth is not infrequently experienced with ependymomas. Isn't there an analogy between ependymoma and pinealoma of ependymal origin? If this supposition may be taken for granted, ectopic histogenesis in my three cases can be coherently explained, i.e. a pinealoma originating from ventricular ependyma has developed extraventricularly in cases 1 and 2, and intraventricularly in case 3.

Summary

1. Three cases of ectopic pinealoma have been reported, case 1 in the left frontal lobe, case 2 in the region of the right basal ganglia and case 3 in the left lateral ventricle.
2. Ependymal origin of the pinealoma seems to be probable.

References