Case Report

Subarachnoid spread of germinoma mimicking tuberculous meningitis

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A case of pineal germinoma spreading along the basal subarachnoid space, clinically and macroscopically at autopsy, resembling tuberculous basal arachnoiditis is reported. Need to carry out CSF cytology of even hemorrhagic CSF is stressed, to diagnose the condition.

Key Words: Germinoma, tuberculous meningitis, basal meningitis

Introduction

Germ cell tumors (GCT) arising primarily in the central nervous system are rare and constitute only a small fraction of all intracranial tumors. Germinomas are the most common of the GCTs with suprasellar being the most common site. Compression of the adjoining neuroanatomical structures by these tumors accounts for their common modes of presentation with features of raised intracranial tension, visual deficits and neuroendocrine symptoms. Though germinomas are notorious for their propensity to spread into the CSF pathways, primary presentation with features of CSF dissemination along the basal cisterns is extremely rare. This case report illustrates one such case wherein the signs and symptoms caused by the dense subarachnoid dissemination of the tumor were so prominent as to mimic a basal meningitis not just clinically but even on gross examination of the brain at postmortem. Only histopathological examination exposed the true neoplastic nature of the lesion and the disease process.

Case Report

A 23-year-old lady presented with a low-grade intermittent fever for the last two months and associated progressive diminshness of vision affecting the right eye more than the left. But over the last ten days she developed headache and vomiting and soon lapsed into a delirious state, talking irrelevantly and failing to recognize her own relatives. On admission, she was found to be pale, withdrawn, listless and disoriented. Physical examination, except for a few firm, non-matted lymphnodes in the submandibular region, was not contributory. Examination of the nervous system showed signs of meningeal irritation. Optic funduscopy revealed primary optic atrophy on the right. Both pupils were equal and reactive to light. No sensory or motor deficits were present. Cranial nerves, except for the left upper motor neuron facial palsy, were unaffected. Visual fields could not be tested as the patient was non-cooperative.

A lumbar puncture drew uniformly blood-stained CSF fluid under raised pressure. The protein content was 70 mg% and sugar 40 mg%. The large number of RBCs precluded a cell count. The lumbar punctures, repeated on two more occasions were again hemorrhagic. The cell count on one occasion was reported to be 20 cells/mm³ (all lymphocytes). In view of hemorrhagic CSF, the possibility of subarachnoid hemorrhage secondary to aneurysmal leak was considered.

A right carotid angiogram with cross compression did not reveal any aneurysm. A couray ventriculogram showed dilatation of the right lateral ventricle. The foramen of Munroe was patent and the dye was seen to fill the aqueduct and the fourth ventricle. But the anterior third ventricle showed filling defects.

The patient's sensorium deteriorated necessitating a ventriculoperitoneal shunt. The ventricular CSF was also blood-stained. Post-procedure, the patient improved but two weeks later she developed pain radiating from the vertex along the shunt tube with extravasation of CSF requiring shunt revision. Subsequently, a diagnostic open biopsy was attempted through a frontal craniotomy. At surgery, both optic nerves and chiasma were noted to be enlarged and entrapped by abnormal looking tissue which was biopsied and sent for histopathological examination. Tissue diagnosis was inconclusive due to inadequate biopsy. Unfortunately, within a couple of hours post surgery, the patient succumbed.

A partial autopsy confined to the removal of the brain was carried out. On external examination, a blood clot was seen covering the site of the burr hole over the right frontal lobe. A striking finding was a thick, dense organized exudate seen filling the chiasmatic, interpeduncular, pontine cisterns along the base of the brain and entrapping the cranial nerves and cerebral vessels (Figure 1a). The optic nerves were embedded within the exudate that was seen to track laterally along both sylvian fissures while posteriorly encasing the brainstem, pons, medulla and cerebellum, obliterating the lateral foramina of the fourth ventricle. Examination of the coronal slices...
revealed hemorrhagic softening of the right caudate nuclei, putamen and the anterior limb of the internal capsules. The ventricular system was dilated. The right frontal horn was filled with blood dots while the left was filled with necrotic material. The ependymal lining of the ventricles was shaggy. A large hemorrhagic mass was noted in the anterior third ventricle, continuous with the basal subarachnoid space (Figure 1b).

Histopathological examination of the sections from the basal ganglia and hypothalamus showed a large tumor mass that was cystic and hemorrhagic. The mass occupied the anterior and middle third of the third ventricle, infiltrating the hypothalamus, columns of fornix and extending anteriorly into the optic chiasma and optic nerve entering the orbital foramina. The ependymal lining of the third ventricle and choroid plexus were bathed in an acute inflammatory exudate. Tumor seedlings were seen in the temporal horn of the lateral ventricle and the head of the caudate nucleus. The optic nerve was enveloped and infiltrated by the tumor, forming segmented cords and islands disrupting the nerve fibers (Figure 2a). The tumor cells were seen spilling into the subarachnoid space to cover the ventral aspect of the brainstem. Tumor implants were identified within the medulla near the floor of the fourth ventricle infiltrating the lower cranial nerve nuclei.

The pineal gland was totally replaced by the tumor leaving behind only the capsule and concentric calcific bodies to identify the gland. Histomorphologically, the tumor was moderately vascular and had the typical biphasic morphology of a germinoma with large round discrete cells amidst a lymphocytic infiltrate (Figure 2b). The cerebral cortex and cerebellum did not have tumor infiltrate though the surface subarachnoid had occasional germinoma cells. The tumor cells were monomorphic, infiltrating into the brain as lobular masses. Some of the parenchymal and subarachnoid vessels had tumor emboli. The tumor was mitotically active. No areas suggestive of syncytiotrophoblastic components were noted.

Discussion

Germinomas are the most common of the intracranial germ cell tumors. At our Institute, among a total of 6353 intracranial tumors seen over the last 15 years, 25 (0.4%) were germ cell tumors (GCTs). Germinomas of the nervous system constituted 72% (18) of GCTs.

The biology and histological spectrum of intracranial GCTs parallel their gonadal counterparts. Similar to all extragonadal GCTs, those occurring in the CNS too favor the midline. The common clinical manifestations caused by these tumors arise from the compression of the surrounding structures such as the aqueduct to cause hydrocephalus, the tectum to produce paralysis of upward gaze and convergence (Parinaud’s syndrome), the optic chiasma producing visual defects or the disruption of the hypothalamic–hypophyseal axis to cause diabetes insipidus/signs of pituitary failure and even precocious puberty. In this case, low-grade fever of long duration, with signs of meningeal irritation, features of raised intracranial tension and primary optic atrophy suggested a chronic basal meningeocerebral process that has entrapped the optic nerves and chiasma. Being a tropical country where tuberculosis is rampant, meningitis of tuberculous etiology figured high on the list. But lumbar puncture done on three occasions was consistently blood-stained. This unfortunately
precluded a cell count but the protein and sugar content were found to be normal. Even an open biopsy was attempted but was inconclusive as it had missed the lesion. Hence, ante-mortem no conclusive diagnosis could be reached, though the possibility of tuberculous basal meningitis (TBM) extending to the anterior third ventricle was entertained. On gross examination of the brain at autopsy, the findings of a thick basal exudate entrapping the cranial nerves and vessels, seemed to confirm the original clinical suspicion of a tuberculous meningitis and the hemorrhagic and softened lesions noted in the right basal ganglia and hypothalamus were interpreted as secondary to tuberculous arteritis. But the histopathological examination revealed the true nature of the disease process to be a pineal germinoma with extensive subarachnoid spread filling the basal cisterns and entrapping the cranial nerves. The marked degree of CSF spread was responsible for the mistaken diagnosis of TBM clinically as well as on gross examination at autopsy. Germinomas are known to invade the subarachnoid space, ventricular cavity, spinal cord and hypothalamus.\textsuperscript{3,4} The incidence of cerebrospinal seedling of the tumor cells has been estimated to range from 7 to 12\% of the cases but higher figures up to 57\% have also been reported in the literature.\textsuperscript{5} The leptomeningeal spread can be easily detected by a simple cytological analysis of the CSF.\textsuperscript{6} This test has assumed great diagnostic importance ever since radiotherapy has become the primary mode of treatment. A positive CSF cytology in suspected cases is considered sufficient to institute radiation therapy even without the necessity of a tissue diagnosis.\textsuperscript{7} With the advent of more advanced imaging modalities such as the CT scan and MRI the chances of missing these lesions are now remote. In this particular case, these facilities were unavailable as it was seen in the pre-CT era.

Interestingly, our case showed direct extension of the tumors into the optic tract causing blindness. Germinoma infiltrating the optic tract is rarely reported in the literature.\textsuperscript{8,9} This case also serves to illustrate the importance of keeping in mind that even tumors can present with features that mimic a basal meningitis. Especially in a tropical country such as ours where tuberculosis is rampant, tumors masquerading as TBM may be missed unless carefully investigated.

References


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