Chiasmatic Syndrome: Clinical Pathological Conference

Jose Bebin
Robert S. Knighton

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Resume of Case (HFH #107 02 16)

This 69-year-old white male was first seen in early 1964 with a complaint of "double vision" of several month's duration. Past health was noncontributory to the presenting complaint.

On examination, his visual complaint was not helped by covering one eye. There was no nystagmus and the external ocular movements were normal. The optic discs were slightly pale. Formal visual field testing revealed a bitemporal hemianopsia. The left face showed some questionable flattening. The right abdominal reflexes were absent. Otherwise, the general and neurological examinations were negative. He related an episode, lasting a day or two, in which his right arm seemed clumsy and he was mildly confused.

On admission, routine blood work, urinalysis and chest x-ray were normal. Plain skull films and laminograms of the sellar area did not show any abnormalities of the sella. However, a faint band of calcification was seen extending superiorly from the sella. Some calcification was seen in the carotid vessels. A right brachial arteriogram showed diffuse atherosclerotic changes but no occlusions. The right vertebral artery was small. The vascular pattern indicated some ventricular dilation. There was no evidence for any abnormal circulation. Pneumoencephalography showed a dilated right lateral ventricle. No air entered the left lateral ventricle. The anterior portion of the third ventricle was not filled and the films suggested the presence of a mass in this area. A brain scan was reported as a class five scan with a heavy area of uptake in the suprasellar region.

*Departments of Pathology, Neurology and Psychiatry.
**Chief, Division of Neurological Surgery.
Progress examinations during the course of these investigations showed a right lower facial weakness and very minimal signs of pyramidal type weakness in the right arm and leg.

A right frontal craniotomy was done and the third ventricle explored. A cystic structure was seen and removed. The pathological report was normal choroid plexus. The day following this procedure, the patient was obtunded and had a left hemiplegia. A further operative procedure was done to drain a small epidural hematoma. No improvement followed and the third ventricle was re-explored and no clot was found. A ventriculostomy was done. No improvement was shown, and four days later, a ventriculocaval shunt was performed. Once again, no change was noted. A week later, a further ventriculostomy was done in the left lateral ventricle. His level of consciousness improved greatly.

Terminally the patient developed a fever of 104° with purulent material suctioned from his trachea. He went into a deep coma and died on the 22nd day after his initial operation.

Discussion

Dr. Knighton:

Today's patient for discussion is a 69-year-old man with a history dating back an undetermined period of time, but at least several months and probably longer. His symptoms and findings are related primarily to the visual system, with a history of double vision of nonspecific nature of several months' duration, and the obvious findings of mild optic atrophy and a symmetrical bitemporal hemianopsia. A mild right hemiparesis and a questionable left facial paresis were also noted. The remainder of his general physical examination and general laboratory studies were noncontributory to the current problem.

We are dealing then with a midline lesion in the region of the optic chiasm, which, as near as I can determine from the history, is progressive. Our differential diagnoses therefore must include the various space-occupying lesions that may be present in the region of the optic chiasm. We must also briefly consider remote lesions which can produce symmetrical enlargement of the ventricular system with dilatation of the third ventricle producing an effect upon the chiasm, but I believe this possibility is so remote in this particular instance that I will say nothing more about it. In fact, we have evidence, as will be pointed out later in the ventricular studies, that the third ventricle was not enlarged. This leaves us with the space-occupying lesions as shown on this table. These are not in order of preference:

1. Colloid cyst of the third ventricle.
2. Chordoma of the clivus.
3. Nasopharyngeal carcinoma.
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5. "Giant" aneurysm.

6. Pituitary adenoma.

7. Craniopharyngioma.

8. Suprasellar meningioma.

I will now consider each of these lesions in reference to the symptoms and findings presented in the protocol.

1. Colloid cyst of the third ventricle. While this tumor might have to be considered in the differential diagnosis, it is extremely rare for such a lesion to produce visual field changes, and specifically so in the absence of symptoms of increased intracranial pressure for which this type of tumor is noted; namely, headache, either intermittent positional, or continuous, and possibly some alteration of consciousness. Likewise, this would definitely have been found on exposure of the third ventricle through the interventricular foramen, as was carried out. I believe, therefore, that this lesion can be eliminated from consideration.

2. Chordomas of the clivus can occasionally produce changes in the visual field, however they are not noted for this. They nearly always involve cranial nerves other than the optic, usually the sixth and as the lesion progresses, the third and fifth, and sometimes the remaining cranial nerves. By the time such a lesion would be large enough to produce visual field changes, which, if present, would likely not be symmetrical, I would expect definite x-ray changes on the submentovertex and perhaps other views of the skull, which were not present in this patient. I think, therefore, that we can also eliminate this lesion from further consideration.

3. Nasopharyngeal carcinoma should be considered in the differential diagnosis, because occasionally it will erode through the floor of the skull and act as a midline mass lesion extending upward into the hypothalamus with pressure and stretching of optic nerves that can produce visual field changes. However, this lesion, as with the chordoma, should show definite evidence of erosion of the base of the skull, and the visual field changes would not likely be as symmetrical as in our patient. I think, therefore, that this lesion may also be eliminated from further consideration.

4. Chiasmal glioma. Gliomas of the optic chiasm produce progressive visual field changes and optic nerve pallor. However, the visual field changes are usually not strictly bitemporal but are more likely to be of the optic nerve type in that they produce scotomatas and progressive loss of acuity. Likewise, they are limited almost entirely to childhood. The patients usually have other stigmata of neurofibromatosis and x-rays of the skull show enlargement of one or the other optic foramen. This diagnosis, therefore, can be eliminated from further consideration.

5. "Giant" aneurysm. Occasionally aneurysms of either the internal carotid artery or the anterior communicating artery can reach tremendous size without rupturing and produce symptoms by direct compression of the optic nerves or chiasm. In other words, they act as an expanding lesion. The aneurysms that arise from the
anterior cerebral or communicating arteries usually do not produce symmetrical visual field changes, but present more with an effect upon one or the other optic nerves and subsequently chiasm, with loss of visual acuity a prominent feature accompanying any visual field loss that might be present. "Giant" aneurysms of the internal carotid artery, however, will often enlarge the sella and produce visual field changes as seen in this patient, and likewise, can also have a calcification in the wall, as in the x-rays described in this patient. However, with an aneurysm of this type large enough to produce visual field changes, other changes in the x-ray should be present. These changes might be the same as are seen in pituitary adenoma, i.e., an intrasellar type of sellar enlargement. There may also be thinning of the anterior clinoid process on one or both sides, and often the superior orbital fissure is widened and occasionally the optic foramen can be slightly enlarged or decalcified, particularly on its medial border. None of these x-ray changes, other than the linear calcification, are described in this patient, and in addition, he had negative arteriograms. I, therefore, feel that while this should be considered in passing, it cannot be the diagnosis in our present case.

6. Pituitary adenoma. The tumor most likely to produce a symmetrical bitemporal hemianopsia is the adenoma of the pituitary gland. Of this group the chromophobe is the most common, but eosinophile or mixed adenomas may produce the same changes. Our protocol does not give us the information as to how the visual field changes progressed, so that we are unable to determine, from the field changes present, whether the lesion began as an infra-chiasmal type, which generally produces a change in the upper temporal quadrants first and then progresses to lower temporal quadrants as the lesion enlarges. This is the characteristic change of the pituitary adenoma, which of course is infra-chiasmal. None of the usual secondary symptoms of hyper- or hypopituitarism are listed, but in a patient of this age, symptoms of hypogonadism or those attributed to hypothyroidism would not be particularly noticeable. Pituitary adenomas can produce transient or permanent hemiparesis when they enlarge sufficiently to distort the brainstem by posterior or lateral extension, so that all of the symptoms and findings present in this particular patient could be accounted for on the basis of such a lesion, with one or two exceptions. First of all, x-rays of the skull are stated to show a normal sella turcica, or at least no evidence of intrasellar enlargement. This, certainly, is not the common finding in such a lesion, which classically gives the intrasellar type of enlargement of the pituitary fossa, with thinning of the posterior clinoid and a double contour appearance to the base of the sella. Rarely, a pituitary tumor can break through the diaphragma sella before sellar enlargement occurs, and this is a possibility in this patient. It can likewise produce a class V scan. Calcification can occur in an adenoma, but is not usually linear. So, while I believe this lesion is probably not the one present in this patient, it cannot be entirely excluded. The normal sella is strong presumptive evidence against it.

This brings us up to our last two choices, the craniopharyngioma and the suprasellar meningioma. I will discuss craniopharyngioma first. This lesion, which is generally thought to arise from a remnant of Rathke’s pouch or Erdheim’s epithelial rests, is usually considered to be a tumor of childhood, although we know that it can present its first clinical manifestations at any time in adult life. In other words,
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these patients are not always the adiposogenital type individual with obvious hypothalamic involvement, and they may lead a perfectly normal existence until progressive enlargement of the tumor is sufficient to produce neurological dysfunction. The slow growth rate can account for an essentially normally functioning hypothalamus and the only presenting symptoms might well be related to the visual system. These tumors in adults differ from the childhood type in that they usually do not present with papilledema, whereas in children this is the usual finding. They do have primary optic atrophy and changes in the visual fields. Calcification is not as prominent in the adult type as it is in childhood, but it may be present. The calcification might be limited to the capsule and present as a linear type of calcification as described in the x-ray findings in this patient. The visual fields are usually not entirely symmetrical in patients with craniopharyngioma, but they can be. In contrast with the pituitary tumor, the craniopharyngioma usually distorts the chiasm from behind. The field loss, theoretically at least, should involve the inferior temporal quadrants first and possibly involve the macular fibers which cross posteriorly, as well. The sella turcica is usually not enlarged but a truncated posterior clinoid process or slightly flattened sella is often seen in this lesion. Neither of these latter changes is mentioned in the protocol. Some degree of hemiparesis would not be unusual in a patient with a craniopharyngioma, so I feel that the findings as listed in the protocol could well fit with the adult type of craniopharyngioma. The only finding against this lesion is the class V brain scan. Generally, the craniopharyngiomas that we have scanned have not shown significant uptake, and a class V, as reported at this hospital, usually indicates dense uptake diagnostic of tumor. I think, therefore, that until I see the radiographic studies listed I will be unable to decide whether this patient’s lesion is a craniophyngioma.

The last differential that I have listed is suprasellar meningioma. In 1930, Harvey Cushing reported 17 operated cases of suprasellar meningioma and characterized the preoperative findings as the “chiasmal syndrome.” He defined the “chiasmal syndrome” as primary optic atrophy with bitemporal field defects in adults showing an essentially normal sella turcica. I believe that our protocol fits this description fairly accurately. However, since this article was written, and with the improvement in x-ray techniques, the suprasellar tumors have been somewhat easier of specific diagnosis. While it is possible not to have changes in the sella with this type of lesion, usually increased calcification due to bone reaction is noted in the region of the tuberculum sella, and usually the bitemporal field defects are not perfectly symmetrical, although they may be. When calcification is present in a suprasellar meningioma, it usually is not a linear calcification, but more likely to radiate out from the site of origin of the lesion. Likewise, an angiogram should reveal a vascular pattern in such a lesion, although it is remotely possible that it would not. Also, since this lesions arises anterior to the chiasm, the anterior cerebral arteries are usually displaced posteriorly by this tumor. A small one would not necessarily show this change. The class V brain scan would be most consistent with meningioma, for this is the one type of tumor that most consistently shows a high uptake of Mercury and Technesium. The changes as reported in the pneumoencephalogram could occur in either pituitary adenoma, craniopharyngioma or suprasellar meningioma, so that this test does not help us in our differential diagnosis.
In summary, we have three lesions which must be considered in this patient: pituitary adenoma, craniopharyngioma, and meningioma, with pituitary adenoma being least likely, because of absence of enlargement of the sella turcica. The two most likely diagnoses are, therefore, craniopharyngioma or suprasellar meningioma. Before making a choice between these, I believe that I would have to see the x-rays for further help.

**X-rays of Skull:**

![Figure 1](image)

Pneumoencephalogram: Rounded filling defect in the anterior third ventricle.

The x-rays of the skull appear essentially normal. The sella is not enlarged. I see no thickening of the bone. The linear calcification mentioned in the report is not too obvious. The arteriogram shows an essentially normal vascular pattern. The brain scan does not appear to be class V in my opinion. The pneumoencephalogram is positive and, I believe, confirms the diagnosis. It reveals a symmetrically rounded filling defect in the anterior third ventricle suggestive of a posterior suprasellar mass. A small nubbin of this can be seen extending downward into the prepontine cistern. The rostral extent is not clear. (Fig. 1)

My final diagnosis, on the basis of his findings and this study, is craniopharyngioma.

**Pathology**

_By Dr. Bebin:_

The most important postmortem findings were in the central nervous system. These will be described in detail. In addition, the autopsy disclosed the following: moderate degree of left ventricular hypertrophy and arteriosclerosis of the coronary arteries with focal calcification. Both lungs were congested, edematous, with focal areas of bronchopneumonia. There was a small splenic infarction and fatty metamorphosis of the liver. The endocrine system was normal.
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The examination of the head showed a right parietal bone flap (6 x 5 x 5 cm) and a right ventriculocaval shunt with a Spitz-Holter valve in place.

The brain weighed 1430 gm. It appeared edematous with prominent bilateral cerebellar pressure cones and moderately congested leptomeninges. The cerebral blood vessels showed a moderate degree of arteriosclerosis.

Over the convexity of the right parietal lobe a 3 x 1 cm surgical wound was present. In the basal aspect of the brain, a coarsely granular and lobular tumor was present. This included the optic chiasma, infundibulum and mammillary regions; laterally it displaced the internal carotid arteries. The pituitary fossa was enlarged and the hypophysis compressed. A midsagittal section of the brain (Fig. 2) disclosed that this tumor was partially cystic containing a granular mucoid material. This neoplasm obliterated the anterior two thirds of the third ventricle and extended upward to the neighborhood of the interventricular foramina. In the rostro caudal direction, it extended from the lamina terminalis to the mammillary region. It measured 3½ x 2½ x 3 cm and it had the typical appearance of a craniopharyngioma.

In addition there was a 2 x 1 cm recent hemorrhage at the operative site. Both lateral ventricles were enlarged, the right more than the left. The brainstem and the cerebellum were edematous and congested. The spinal cord was covered by a recent subarachnoid hemorrhage.

The tumor showed an admixture of cystic and solid areas (Fig. 3). The cyst wall consisted of a squamous epithelium, single-layered and supported by collagenous basal
membrane which separated it from the nervous tissue (hypothalamus). In areas, this epithelium showed evidence of retrogressive changes and formation of microcysts. The cyst contained an abundant amorphus eosinophilic material, cellular debris and cholesterol.

Figure 3
Section of the tumor: solid and cystic craniopharyngioma. H.E. x5

The solid portions of the neoplasm were made up of islands of squamous epithelial cells (resembling the basal layer of the skin) with oval deeply stained nuclei and eosinophilic cytoplasm. The cellular structures were supported by a loose, scanty cellular vascular connective tissue stroma. The most superficial layer of the epithelium appeared columnar. (Fig. 4) This squamous epithelial appearance was more noticeable in the best-preserved areas, where intracellular bridges were present. The central portion showed extensive retrogressive changes, leading to formation of small and large cysts filled with cellular debris and eosinophilic material. In other areas, there was accumulation of lamellar masses of keratin and calcium deposits. (Fig. 5) An inflammatory lymphocytic reaction was present in the degenerative areas.

Both optic nerves and chiasma showed partial demyelination and gliosis. The surrounding brain tissue showed a considerable degree of gliosis and often finger-like extrusions of the neoplasm invaded it. At the limit between the two tissues there were abundant lymphocytic cells present. In addition there were some arteriosclerotic changes of the brain with mineralization of the vessel walls in the globus pallidus.
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Figure 4
Craniopharyngioma: columnar epithelium with sparsely cellular stroma. H.E. x780

Figure 5
Craniopharyngioma showing microcystic changes in various stages and laminated masses of keratin. H.E. x150
Patho-anatomical diagnoses:
   Postoperative edema and congestion.
2. Cerebral arteriosclerosis.
3. Acute bronchopneumonia, bilateral.

Comments

Craniopharyngiomas, in general, are considered to be tumors of children and young adults. The finding of a tumor of this type in an older patient is a rarity. However, the review of the literature on this subject revealed that an appreciable number of patients have their first symptoms after the age of 40, and that differences exist between these and most young patients.

In children there are two main syndromes: (1) visual failure from compression of the optic nerve or chiasma, and (2) symptoms and signs of increased intracranial pressure resulting from obstruction of the third ventricle by the tumor, such as papilledema, headaches, cranial suture separation without specific visual field disturbances.

In adults, visual failure from chiasmal compression remains common and mental disturbances increasingly important while there is a decrease in endocrine manifestations and less tendency by the tumor to obstruct the flow of cerebro-spinal fluid.

Since craniopharyngiomas are congenital, it is not surprising that the majority should present early in life. Their presence in adults suggests that they remain clinically silent until middle life or later and that their rate of growth must be very slow or intermittent.

Bailey et al. (1939) gave the following age distribution in 138 cases: 66 cases under the age of 20; 47 cases between 20 and 40 years of age, and 25 cases older than 40 years of age. Northfield (1957) reviewed 49 cases of craniopharyngiomas. Of them, 13 occurred in patients over 40 years of age; and Ross Russell and Pennybacker (1961) described the clinical manifestations of 24 middle-aged and elderly patients with craniopharyngiomas in whom fluctuant reduction in visual acuity or abnormalities of visual fields were a common and striking feature. Mental changes were noted in half of the patients and in some these changes were the dominant features.

The radiological aspects of craniopharyngiomas are discussed by Barnett (1959). The sella turcica usually was not enlarged. In one half of 62 patients, the anterior clinoid processes were eroded and appeared “pointed,” and the dorsum of the sella and posterior clinoid processes were most affected by the erosion. Abnormal calcific deposits in and about the sella are important and diagnostic. Seventy-five percent of cases show calcification in the form of soft flocculant and irregular densities in the suprasellar area. By pneumonencephalography or ventriculography, 95% had dis-
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tortion of the third ventricle, usually compression exerted from below causing a smooth rounded defect in the anterior portion of the third ventricle. The lateral ventricles were enlarged in 27 patients.

The craniopharyngiomas may be solid or cystic tumors, anatomically related to the pituitary gland and its stalk. They are believed to arise from embryonic nests of squamous epithelium in the pars tuberalis and structurally closely related to the epidermoid cysts of other sites. The truly congenital nature of the epidermoid nests found in the pituitary gland has been debated. Luse and Kernohan (1955) considered them derived from metaplasia of pituitary cells. Craniopharyngiomas constituted about 3% of all intracranial tumors, and more than half occurred in childhood and adolescence (Russell and Rubinstein, 1963). They can develop in fetal life and they are not so rare in later life as the case presented today and others in the literature indicate. The gross and microscopic appearances of these tumors are variable and the case discussed illustrated very well most of the pathological features of this neoplasm.

REFERENCES
