The Empty Sella Syndrome*

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Pituitary tumors often encroach upon and compress the nearby optic nerves and chiasm causing a variety of visual deficits. Chamlin, et al.,1 reported visual field defects in 86%, optic atrophy in 50%, and impaired visual acuity in 32% of these patients with neoplasms of the pituitary. Since these patients may become totally blind if not treated, the primary objective of treatment is to relieve the pressure on the visual pathways by operative decompression, irradiation, separately or in combination. The success of treatment is measured by the degree of improvement in visual function that can be objectively determined and recorded.

Deterioration in visual function after treatment is usually assumed to indicate recurrence of tumor. However, on rare occasions, no tumor is found when the enlarged sella is explored surgically. This condition has been called the "empty sella syndrome,"2 and is a rare complication. We are reporting three cases of this syndrome encountered at the University of California San Francisco Medical Center.

Case Reports

Case 1. A 56-year-old woman was hospitalized in January, 1949, because of a 6-month history of headache and blurring vision without diplopia.

Examination. Upon admission examination disclosed a left temporal hemianopsia and a right upper temporal quadrant defect. Visual acuity on the right was 20/200 and on the left was 20/100. Both optic discs were pale. X-rays of the skull demonstrated an enlarged sella.

First operation. On January 22, 1949, a right frontal craniotomy revealed a large cystic chromophobe adenoma, which was partially removed. The patient received postoperative radiation in a total dose of 3600 R at midplane through two lateral portals.

Initially there was some postoperative improvement in the right temporal visual field; visual acuity also improved to 20/40 on the right and 20/100 on the left (Fig. 1). However, in November, 1950, visual acuity had deteriorated to 20/70 on the right and 20/200 on the left. A large right central scotoma appeared in the right field. There was no further progression of visual loss until July, 1952, when examination disclosed further loss of vision in the right nasal field.

Second operation. An exploratory operation in July, 1952, showed that both optic nerves were white and thin and surrounded by arachnoidal adhesions. The right optic tract appeared normal. No tumor was found in the sella.

Postoperatively, vision in the right eye continued to deteriorate. The patient saw hand motion with her right eye in November, 1952, and had only light perception in September, 1953. The condition of her left eye remained stable with a visual acuity of 20/200 and a total temporal hemianopsia. The patient has not returned for follow-up examination since 1954.

Case 2. This 58-year-old woman, first seen in 1964, had a history of acromegaly of more than 20 years’ duration. In 1949 she had had a course of x-ray therapy to the pituitary, the dosage being unknown. Because of failing vision, she subsequently had received an additional 4317 R in 1962 and 1740 R in 1963.

Examination. At examination in 1964, the patient had only faint light perception in the left eye, and acuity in the right eye was limited to finger counting only. There was a complete right temporal hemianopsia. Data from previous eye examinations were not available. Fundusscopic examination revealed bilateral optic atrophy. She had the typical features of acromegaly. The sella was enlarged and its floor depressed (Fig. 2). Left carotid angiography showed the left internal carotid artery

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displaced laterally and the carotid siphon distorted (Fig. 3). A pneumoencephalogram showed a slightly enlarged left temporal horn and a possibly elevated anterior portion of the third ventricle.

Operation. A left frontotemporal craniotomy was done on July 14, 1964. Marked atrophic changes were seen in the left frontal and temporal lobes. The medial portion of the left lesser sphenoid wing and left anterior clinoid process had been destroyed. The large sella was empty; there was no evidence of tumor. The left optic nerve sagged into the ballooned sella; the right optic nerve was not visualized. On reviewing the pneumoencephalograms, there seemed to be some air in the sella, which had not been noted preoperatively.

After operation there was no improvement in visual function. The patient has been
followed by her private physician who reported in December, 1966, that her vision has remained about the same.

Case 3. A 20-year-old woman was seen in January, 1961, because of poor vision of 1 year's duration. The patient had become amenorrheic 4 months before admission and then developed a bitemporal hemianopsia and retro-orbital headache just before admission.

Examination. Visual acuity of the left eye was 20/200, that of the right 20/30. The sella was enlarged. Carotid arteriogram showed uncoiling of the siphon and slight elevation of the anterior cerebral artery (Figs. 4 and 5).

First operation. On January 5, 1961, a right frontal craniotomy was done. A large, soft intrasellar chromphobe adenoma was found and removed subtotaly with only a small piece of capsule being left on the left optic nerve and chiasm. Postoperative radiation therapy was given, the tumor dose being 3600 R.

Some recovery of visual fields on both sides occurred (Fig. 6). The patient was followed at home by her family physician and then by the physician at the college she attended. In January, 1963, her family physician reported that her visual fields were normal.

Second examination. In December, 1966, the patient returned to our service with complaints of frontal headaches and blurred vision of 2 months' duration. On examination, she again had a dense (3/1000) temporal hemianopsia on the left and a superior temporal quadranopsia on the right. Visual acuity was 20/30 in the right eye and 20/200 in the left.

She was readmitted for further operation. A repeat pneumoencephalogram showed the sella filled with air (Fig. 7). We cancelled the operation and are following the patient very closely to detect any further visual deterioration. As of March, 1967, there has been none.

Discussion

The "empty sella syndrome" is a poorly defined term. It is used verbally quite often, but scarcely anything has been written about it. The only direct reference to it is in a case described by Colby and Kearns\textsuperscript{2} from the
Fig. 4. Right carotid arteriogram showing displacement of carotid siphon and anterior cerebral artery (Case 3).

Fig. 5. Left carotid arteriogram showing displacement of carotid siphon and anterior cerebral artery (Case 3).
Mayo Clinic in a review of their results of radiation therapy of 149 pituitary tumors. They wrote: "A rather rare complication of irradiation is the 'empty sella syndrome,' in which a patient, treated a number of years previously with improvement in vision, later experiences visual impairment and is operated upon with the presumption that his tumor is growing again. At operation no tumor is found." This, of course, is very much like our Case 2.

Similar cases of delayed visual deterioration have been reported by others, including Morello and Frera, Walsh, and Hartog, et al. In none of the these cases could deterioration of visual function be attributed to a recurrent tumor. Therefore, some other explanation must be sought.

All three of our patients received radiation treatment at various times in the course of their disease. Most recent reports have indicated that the central nervous system can tolerate up to 5000 R with modern techniques. Necrosis of the calvarium and various parts of the brain has been recorded after excessive doses of irradiation.

Only our patient in Case 2 received an excessive dose. The first course of irradiation was given in 1949 and the exact dose was unknown. With visual deterioration, an additional 6057 R were delivered. Therefore, the total sum would certainly have been higher than the accepted safe limit. At the time of operation, marked destructive changes were seen in the sphenoid wing and clinoid process, and the frontal and temporal lobes were atrophic. The left optic nerve was white and 

FIG. 6. Postoperative serial visual field examinations showing early improvement and late recurrence of field defect (Case 3).

FIG. 7. Pneumoencephalogram (midline tomogram) showing air in the interpeduncular system as well as in the sella. Third ventricle is faintly seen (Case 3).
atrophic on gross inspection. It is conceivable that this patient’s loss of vision was directly due to radiation damage.

A second possible explanation exists. At the time of operation the optic nerve was seen dipping into the enlarged and yet empty sella. The optic nerve and chiasm in the patient described by Colby and Kearns had also been drawn into the sella, apparently by scar tissue; this was considered by the authors to be the cause of their patient’s recurrent visual disturbance. The optic nerves of the patients described by Hartog, et al., also sagged into the empty sella. It is conceivable that the traction and distortion of the nerve and the compression against the underlying bone could cause the visual deterioration.

Two of our patients had initial surgical decompression preceding irradiation. It is possible that postoperative changes had, in some fashion, enhanced the effect of irradiation. The patient in Case I underwent a second exploration because of progressive visual loss. No tumor was seen at that time, but numerous adhesions were found. It has been well documented that progressive blindness may result from: 1) adhesive arachnoiditis around the optic nerves and chiasm following trauma, hemorrhage, or meningitis, and 2) diffuse meningeal carcinomatosis.

In these cases, visual loss is usually said to be caused by vascular strangulation. One of the well-known changes following irradiation is vasculitis and occlusion of small vessels. Therefore, the postoperative adhesions and postirradiation vasculitis may combine to cause a delayed necrosis and atrophy of the optic nerve and chiasm.

Case 3 is interesting because of the appearance of the air-filled sella in the pneumoencephalogram. Robertson, in his textbook, described a large arachnoid cyst in the sella which stretched the optic nerves to cause bitemporal hemianopsia. On exploration the sella was found to be enlarged and the cyst filled with clear cerebrospinal fluid; no tumor or solid tissue was seen. In our case, the air entered and left the sella freely with change in head position; thus, it is unlikely that our patient had a true arachnoid cyst. It is difficult to conceive of a cyst that has free communication with the cistern, yet is able to exert significant pressure on the optic structures. Ring and Waddington recently reported two patients in whom the sella was filled by air on pneumoencephalography. These patients had no visual symptoms or signs and neither was explored.

**Summary**

We have reported three cases in which patients with pituitary tumors treated by surgery or x-ray later developed visual dysfunction without gross evidence of tumor recurrence. We have emphasized the appropriateness of the term “empty sella syndrome” for this complication and have suggested various explanations to account for the pathogenesis of this rather uncommon condition.

**References**


7. Walsh, F. B. Personal communication.