Post-irradiation pneumatocele in a massive chromophobe adenoma

Noble J. David, James L. Poppen, and Fredie Gargano

From the Departments of Neurology, Ophthalmology and Radiology, University of Miami, and the Lahey Clinic, Boston, Massachusetts, U.S.A.

Numerous previous reports have emphasized the capricious behaviour of the chromophobe adenoma, commonest of the intrasellar neoplasms. Their unpredictable size and direction of growth, as well as their puzzling clinical manifestations when complicated by 'pituitary apoplexy', have been the subject of excellent descriptions—among others, those of Jefferson (1940) and Walsh (1949, 1957). The tendency to form massive subfrontal extensions which damage the anterior visual pathways is quite characteristic of this tumour, as is visual improvement after radiation therapy. In the patient to be described, air filled the necrotic tumour cyst following this form of therapy, an event for which we can find no precedent in the literature.

Case Report

A 64-year-old retired schoolteacher was referred to the Bascom Palmer Eye Institute on 25 August 1965 for evaluation of impaired vision of four years' duration. She described the insidious onset of a mild and gradually progressive impairment of leftward vision, noted mostly while reading or driving. She believed there had been definite worsening in the two months before examination, in that she read 'pieces of words' and had to sweep her gaze far to the left to pick up phrases and lines. She limited her driving to an absolute minimum because she tended not to notice cars coming from the left'. No other neurological symptoms were described. She had observed no change in weight, appearance, or voice and had noted no other systemic symptoms, including temperature intolerance. She had suffered no headaches or any intellectual or emotional changes.

Ophthalmological examination revealed visual fields (Fig. 1a) indicating an incongruous homonymous left hemianopsia indicative of right optic tract involvement with some chiasmal damage. Central visual acuity was 20/20 in the right eye, 20/50 in the left. The only funduscopic abnormality was questionable optic atrophy in the left eye. Except for the field defect, neurological evaluation was entirely within normal limits. Olfaction was intact to coffee and soap, cranial nerves III through XII were normal. Her affect, memory, orientation, and intellect were intact. No abnormal reflexes were found and the remainder of the general and neurological examinations were unremarkable.

Routine skull films (Fig. 2) revealed marked enlargement of the sella turcica with undercutting of both anterior clinoids and erosion of both posterior clinoid processes. Destruction of the floor of the sella with marked erosion of the sphenoid sinus had occurred.

The patient entered the hospital on 29 August 1965 for further studies. Routine laboratory data including haemogram, urinalysis, serum electrolytes, and fasting blood sugar were all normal. Other tests showed VDR non-reactive, PBI 4·8, spinal fluid clear, colourless, with 1 white cell/cu.mm, 9 red cells/cu.mm, protein 54, serology negative. Twenty-four hour urinary excretion of 17-ketosteroids varied from 5·2 to 9·1 and 17hydroxycorticoids from 2·0 to 4·1, both values ranging within the low side of normal. Thyroid stimulating hormone was less than one mouse unit in 24 hr. Radioactive iodine uptake by the thyroid gland was 27% in the 24 hr. Radioactive brain scanning revealed a midline density 4 cm in diameter in the floor of the frontal fossa contiguous in its anatomical shadow with the region of the sella turcica. Electroencephalography was within normal limits. Pneumoencephalography was combined with bilateral carotid arteriography in studying this mass (Fig. 3a and b). The angiograms demonstrated lateral displacement of both internal carotid arteries from the intracavernous portions to the bifurcations. Both posterior communicating arteries and anterior choroidal arteries were laterally displaced. Both anterior cerebral arteries were elevated with stretching of both frontal polar branches. An unrelated finding was the demonstration of a saccular aneurysm, 1/2 x cm, at the bifurcation of the right middle cerebral artery. The venous phase angiograms revealed lateral and upward displacement of the anterior portion of the basal vein of Rosenthal. The septal veins and deep central veins were displaced dorsally. The pneumoencephalograms showed a midline mass which elevated the floor of both anterior horns to the lateral ventricles. The anterior recesses of the third ventricle were amputated by this mass. A primary pituitary tumour was considered the most likely diagnostic possibility.

On 18 September 1965 the patient was transferred to the Lahey Clinic for further care. Because she felt well and wished to avoid surgery if possible, it was decided to attempt initial treatment with irradiation. In the following month she received 4000 r to the pituitary area through a 5 cm diameter port (two million volts...
Post-irradiation pneumatocele in a massive chromophobe adenoma

FIG. 1a. 17 September 1965. Visual fields at the time of first examination. A left homonymous hemianopsia is combined with right temporal involvement, interpreted as a combination of right optic tract and chiasmal damage. (Left eye: vision 20/40; test objects BL=18/1000w, 11/=6/1000w. Right eye: vision 20/20; test objects BL=18/1000w, 11/=6/1000w.)

FIG. 1b. 3 November 1965. Visual fields at the termination of one month of irradiation therapy to the tumour. A mild upper bitemporal quadrantanopsia with impairment of visual function, improvement sustained for the next 10 months. (Left eye: vision 20/40; test objects 8/1000 PL. Right eye: vision 20/30; test objects 8/1000 PL.)

FIG. 1c. 31 August 1966. Rapid loss of visual acuity and deterioration of fields coincident with the discovery of the pneumatocele within the tumour. Two weeks earlier her visual fields were identical with those in Fig. 1b. (Left eye: vision 20/80; test objects 8/1000 PL. Right eye: vision 20/200; test objects 8/1000 PL.)

Technique with 360 degrees rotation). On 12 October 1965, two months after the completion of treatment, visual field examination revealed considerable improvement (Fig. 1b) with no significant field defect on the right and great reduction in the homonymous left-sided field encroachment.

Throughout the first seven months of 1966 she kept a mild upper temporal defect bilaterally with some suggestion of nasal involvement. Visual acuity approached 20/20 in both eyes on repeated tests. She read several books weekly, drove her car, and was entirely asymptomatic. Early in August 1966, she began to experience bilateral retro-orbital, frontal, and temporal headaches which were variable in intensity, sharp, and stabbing, and reminded her of her lifelong 'sinus headaches', made worse by jarring footsteps and nose-blowing. She had noted no rhinorrhea, fever, stiff neck, or change in mentation. She thought her vision had 'fogged a bit' and described the peculiar sensation of thinking that everything looked 'bright' to her. On 31 August 1966 she complained that her vision was definitely worse and on examination was found to have an acuity in the right eye of 20/200, left eye 20/80. Field examination (Fig. 1c) revealed a marked increase in her homonymous hemianopsia with a dense bitemporal upper quadratic field loss. She was readmitted to the hospital on 29 August 1966 at which time routine skull radiographs revealed a remarkable air shadow filling the anticipated internal contours of the tumour (Fig. 4a and b). The pneumatocele was bilobate in frontal projection and about 2½ cm in diameter in the lateral projection, in which view a double shadow could be seen. Fifteen cubic centimetres of air were introduced by lumbar puncture to show the tumour's relationship to the ventricles (Fig. 5). The lateral ventricles were pushed upward, the third ventricle backward by this mass. Cerebrospinal fluid con-
tained 1 white cell and a protein content of 45 mg%. No cerebrospinal rhinorrhea was detected at any time and the patient was afebrile. Prompt surgical intervention was advised; the patient boarded a commercial airliner for Boston, Mass., to enter the Lahey Clinic. After ascent, she developed the severest frontal head pain she had ever known. She described a virtually ‘blinding headache’, remarking that her companion’s magazine looked hazy and dark, and inquiring if the lights in the cabin had been turned off. Immediately thereafter she became intensely nauseated and vomited repeatedly. Of the succeeding hours and days, she had few distinct memories except for head pain. She said of her flight, ‘I guess that air in my head expanded just like a balloon.’ Her physicians could find little fault with this hypothesis.

She was admitted to the Baptist Hospital in Boston where, six days later, right frontal craniotomy was performed. The right optic nerve was compressed at the chiasmal level; the right superior orbital plate and ethmoid sinuses had been opened by tumour invasion. Tumour was wrapped around the right optic nerve. After its removal one could see pulsation of the spinal fluid within the subarachnoid space surrounding the proximal portion of the optic nerve. A large cavernous opening into the sphenoid sinus was discovered, which extended forward into the region of the paranasal sinuses. The tumour was excised with exception of a few fragments of capsule. Exploration revealed that the tumour, which was certainly made necrotic and cystic by radiation therapy, had grown forward and upward from the pituitary sella, surrounding and invading the sphenoid sinus and para-optic nerve area inferiorly. The right middle cerebral aneurysm was not explored. Flattened temporalis muscle was used to seal off the cranial defect through which the tumour had invaded the sphenoid and ethmoid sinuses.

Post-operatively, the patient’s visual fields rapidly returned to their best post-irradiation function. The patient had a left 6th nerve palsy which cleared within six months after the operation. Three months after the operation, the patient began to notice crystalline clear fluid draining from the nostrils when she bent over or strained, and nightly wetting of her pillow from this.

FIG. 3a and b. Anterior-posterior and lateral views of arterial phase of right carotid arteriogram. The films demonstrate the area of subfrontal extension. The anterior cerebral artery and frontal polar branch are draped over the tumour. The right middle cerebral aneurysm incidentally found is also clearly demonstrated.
Post-irradiation pneumatocele in a massive chromophobe adenoma

FIG. 4a and b. Routine PA and lateral skull film showing air-filled tumour capsule. Residual tumour is seen at sellar level.

colourless nasal discharge. The rhinorrhea lasted about three weeks and, untreated, it gradually dried up. She remained asymptomatic with good bilateral visual function.

DISCUSSION

This patient clearly illustrates the enormous size subfrontal masses may obtain with no detectable cerebral defect, save for involvement of sight. Operative decompression was initially deferred because of the patient's age and strong aversion to surgery, the presumptive diagnosis of chromophobe adenoma being practically established by her salutary response to irradiation. The spectacular pneumatocele which subsequently developed and rapidly enlarged causing headaches and visual deterioration is unique, to the best of our knowledge. Operation revealed the source of air to be a defect through the sphenoid sinus, the floor of which had been eroded by the tumour. Further bone necrosis may have been caused by irradiation. This source of air communicated with the tumour cyst but not with the cerebrospinal subarachnoid space before operative removal. Her sudden deterioration in a commercial jet airplane after cabin decompression would indicate that the mass of air either was temporarily sealed off or had an effective valve preventing its outflow. In spite of this misadventure, surgical decompression was probably made easier by relaxing this cystic subfrontal mass before its removal. The patient did not remember expelling any necrotic material as nasal discharge that might have represented fragments of the degenerating chromophobe adenoma.

SUMMARY

Irradiation of the large subfrontal extension of a chromophobe adenoma resulted in initial improvement followed by the development of a large pneumatocele filling the internal contours of this tumour. Before its surgical excision, this intra-

FIG. 5. Lateral pneumoencephalogram film demonstrating relationship of air-filled tumour bed to frontal horns, third ventricle, and basal cisterns.
cranial balloon produced rapid worsening of the patient's symptoms, greatly aggravated by an airplane flight. On surgical removal, the histopathology of chromophobe adenoma was proven and visual function returned to its best pre-operative levels.

REFERENCES