Retrobulbar Orbital Myxoma and its Detection by Ultrasonography

Case Report

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There have been only six reported cases of orbital myxoma. The additional case we are reporting is particularly interesting because of the role which ultrasonography played in its detection, in the selection of the operative approach, and in the postoperative course.

Case Report

A 31-year-old man from New Orleans, Louisiana, was transferred to the Bronx Veterans Administration Hospital on March 4, 1968, for ultrasonographic examination. Repeated ophthalmological, medical, and roentgenological studies during the previous 2 years had failed to reveal the cause of the protrusion of his right eye.

History. The patient first noted the bulging of his right eye in a photograph taken in November, 1960. It gradually became worse, yet neither pain, visual impairment, nor double vision developed. In early 1961, he consulted an ophthalmologist and continued under his observation until transfer to our hospital.

Received for publication June 3, 1966.

Preoperative Examination. The medical and x-ray examinations again revealed no abnormalities except for the ophthalmological findings. The right eye was markedly protruded and was moderately firm when pressed. The right eye was 5 mm lower than the left (Fig. 1). Hertel exophthalmometer readings were 23 mm/100 mm on the right and 17 mm/100 mm on the left. Schiötz tonometry values were normal. Visual acuity was 20/20 in each eye. The fundoscopic examination revealed only an absent venous pulsation on the right. The extraocular movements were normal and there was no diplopia. All other ophthalmological studies were normal.

A review of photographs taken during adolescence indicated that the protrusion had existed, although to a lesser degree, more than 10 years before admission.

Ultrasonographic Examination. The ultrasonogram revealed a huge saddle-shaped mass astride the right optic nerve. The tumor extended approximately 4.5 cm in an anterior-posterior direction (Figs. 2 and 3). Because of the size, shape, and location of the mass outlined by ultrasonography, it became obvious that its complete removal could only be safely carried out by a transfrontal craniotomy.

Fig. 1. Appearance of the patient 1 week before operation.
patches from fibrous of roughly the pothermic present cm nerve impression tumor removed. By a palpebrae, was WFlo. operation. FIG. 4. Photograph of the tumor specimen.

Fig. 3. Composite drawing derived from the serial ultrasonograms illustrates the size and location of the tumor.

Operation. On April 1, 1963, a right frontal craniotomy was carried out under general endotracheal and hypothermic anesthesia. The paper-thin orbital roof was removed. The optic foramen was also unroofed, exposing the optic nerve. Following incision of the periorbital membrane, orbital fat began to bulge through the opening. After retraction of the frontal nerve and the levator palpebrae, blunt dissection exposed the upper surface of a bluish-gray, partially-hard, nodular, lobulated mass. By severing fine strands and sheaths of connective tissue, it became possible to lift out most of the large tumor mass. A small piece directly overlying the optic nerve was later removed by sharp dissection. It was the impression of the neurosurgeon and ophthalmologist present that the tumor had been totally removed (Fig. 4).

Gross Pathological Examination. The specimen was a roughly ovid piece of gelatinous tissue with strands of fibrous tissue over the surface. It measured $5 \times 2.9 \times 1.8$ cm and weighed 10.6 gm. Gelatinous material exuded from the surface, which was gray and scattered with patches of red and had small nodules which measured up to 0.6 cm in diameter. On section, the interior was similar to the external surface. There was a pseudo-capsule over approximately one half of the lesion; the rest of the surface was bare.

Microscopic Examination. The tumor was composed of loosely-arranged spindle-shaped cells generally oriented in one direction and lying in an edematous connective tissue stroma (Fig. 5). The cells while varying considerably in size, were mostly quite small with dark-staining elliptical, centrally-located nuclei, and cytoplasmic prolongations at one or both ends. Other larger tumor cells showed round nuclei and blunted cytoplasmic ends. Occasionally nucleoli and cytoplasmic vacuoles were seen. In a few instances the tumor cells were bizarre, presenting large oval or round hyperchromatic nuclei. Mitoses were not seen despite the atypical nuclei. The connective tissue between the tumor cells was very loose-textured, with numerous large spaces that looked as if they had been filled with a fluid. Period-
Ultrasonography Detection of Orbital Myxoma

Fig. 6. Ultrasonogram on the 17th postoperative day (above) displays an abnormal area indicated by the arrow. This area is characterized by fewer and weaker echoes than are present in other parts of the orbital fat. Note the similarity to the appearance of the tumor tissue in Fig. 2. The finding was initially interpreted as a possible remnant of the tumor. An ultrasonogram 6 months later (below) showed that this area had diminished in size. Because of the slow growth of the tumor, many years will be required to tell whether this area is tumor remnant or scar tissue.

Fig. 5. Photomicrograph (high power, X950).

ic acid-Schiff stains showed no positive material. The microscopic diagnosis was myxoma (Drs. B. S. Gordon and H. M. Zimmerman).

Postoperative Course. The patient tolerated the procedure well and the wound healed normally. An ultrasonogram done on the first postoperative day raised the possibility that a portion of the tumor approximately 5 mm in diameter still remained in the superior temporal quadrant of the orbit (Fig. 6). The interpretation of this finding is still questionable, not only because of the operative findings and the thorough inspection of the
Myxomas are said to arise from embryonic, muciform tissue, which is the parent substance of several varieties of connective tissue. The occurrence of true myxomas in the orbit is exceptionally rare. In the seven known cases, the tumors were loosely attached to the adjacent structures by connective tissue strands. In only one instance\(^5,9\) were multiple tumors observed. All other reports mention solitary tumors, variously described as soft, elastic, semi-solid, lobulated masses. Although occasionally described as "small," they are usually "large."

The tumors occurred in four females and in three males between the ages of 16 to 56 years. The length of the illness before the first examination varied from 3 to 33 years in five instances.

When the tumors were anterior under the conjunctiva,\(^1,10\) under the upper lid, or under the upper rim of the orbit, they could be both seen and felt.\(^5,6,8,11\) Displacement of the bulbus oculi was observed in five patients including our own,\(^5,6,8,10\) exophthalmos, of marked degree, in four patients,\(^5,7,9,15\) limitations of eye movements in three patients,\(^5,8,10\) and diplopia in two.\(^6,10\) Visual impairment was attributed in one instance to papil-

Fig. 7. Ultrasonogram of the normal left eye at a comparable level of scan shows no such abnormality as that seen in Fig. 6.

area after removal of the tumor, but also because a subsequent ultrasonogram 6 months postoperatively revealed that the suspicious area had diminished in size. An ultrasonogram of the normal left eye is shown in Fig. 7 for comparison. Since this was obviously a slow-growing tumor, a long period of observation will be required to tell whether the area represents tumor remnant or scar tissue.

On April 29, 1963, the patient was discharged. By then the exophthalmos had disappeared (Fig. 8). Visual acuity was normal in both eyes, and there was no limitation of ocular movements. The pupils were equal and reacted briskly to light and near vision.

The patient returned for re-evaluation on October 8-9, 1963. Except for a slight widening of the right palpebral fissure, no evidence of impairment of the visual system was found. The ultrasonographic findings at this time have already been described (Fig. 6).

The patient was re-examined on August 17, 1966. There were no subjective complaints. There was exophthalmos (1 mm) on the right, and diplopia on red glass tests in the primary position and on right lateral gaze. The ultrasonogram showed an increase in the size of the lesion. These findings, however, were not considered significant enough to reach a conclusion regarding recurrence of the tumor.

Discussion

Myxomas are said to arise from embryonic, muciform tissue, which is the parent substance of several varieties of connective tissue. The occurrence of true myxomas in the orbit is exceptionally rare. In the seven known cases, the tumors were loosely attached to the adjacent structures by connective tissue strands. In only one instance\(^5,9\) were multiple tumors observed. All other reports mention solitary tumors, variously described as soft, elastic, semi-solid, lobulated masses. Although occasionally described as "small," they are usually "large."

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Fig. 8. Appearance of the patient 32 days after the operation.
Ultrasoundography Detection of Orbital Myxoma

ledema of 6 diopters, in another to optic atrophy, and in a third to increasing hypermetropia. Pain in the eye was experienced by two patients while awareness of a "foreign body" on lid closure was mentioned by one.

Most myxomas have been removed in one piece. A recurrence of tumor growth was reported only in the case of multiple orbital myxomas; this was not attributed to malignancy, but to incomplete previous removal. The postoperative results usually were characterized by a gratifying disappearance of symptoms, except for those involving severe, preoperative, visual impairment.

X-ray was diagnostically helpful in only one case, where it demonstrated an enlargement of the orbit with obliteration of ipsilateral ethmoid cells, and opacity and haziness of the ipsilateral frontal sinus.

In our case, ultrasonography was extremely helpful. Myxomas possess an acoustic impedance which differs greatly from that of the surrounding tissue. The capsule of the tumor gave very dense, irregular echoes, unlike the uniform echoes of normal orbital fat (Fig. 7). The tumor is easier to observe on the undifferentiated film than on the differentiated film, which implies a gradual rather than an abrupt change in acoustic impedance at the tumor tissue interface.

The use of ultrasonography for the localization of orbital lesions has been reported in previous publications. The ultrasonic principle of a pulse echo is the same as that of the simple ultrasoundoscope used in the more familiar identification of midline structures of the brain. In that device, a transducer is held in the hand and the returning echoes are displayed as a graph on the face of the oscilloscope. The displacement from the base line equals the strength of the echo and the distance between the echoes represent intracerebral distance.

In our more sophisticated ultrasonoscope, the pulses are represented as points of light of varying intensity rather than displacements from the baseline of the oscilloscope. In addition, scanning is performed mechanically and the oscilloscope trace line is shifted in direct proportion to the motion of the transducer. The resulting image formed on the face of the cathode ray tube is a tomograph of the tissues examined. The ultrasonogram is the photograph of the image on the face of the cathode ray tube. Vertical tissue dimension is determined by serial ultrasonography which is produced by recording succeeding scans at successive levels of examination.

In a series of 54 cases of unilateral exophthalmos, ultrasonography localized orbital tumors four times more frequently than x-ray examination. These results may be biased because the patients were generally referred to us following a negative roentgenological examination.

Summary

We have reported the successful removal of a large retrobulbar myxoma through a transfrontal craniotomy. Ultrasonography played an important part in the detection and delineation of this tumor in the selection of the operative approach, and in postoperative evaluation.

References