

TUMORS OF THE ORBIT

L. DOCTER, M.D.,* and ROSCOE J. KENNEDY, M.D.

Department of Ophthalmology

TUMORS of the orbit are not uncommon and must be suspected in every patient with exophthalmos, especially if the condition is unilateral. Benign tumors are the most frequent and include hemangioma, pseudotumor, meningioma, dermoid cyst, osteoma, neuroma, and lipoma. The malignant growths are sarcoma, carcinoma (frequently of the lacrimal gland), lymphoma, leukemic infiltrations, and tumors of the optic nerve.

Clinical Manifestations

The most consistent finding in patients with orbital tumors is exophthalmos, which may be a straight forward protrusion if the growth arises within the muscle cone of the eye or a lateral or vertical displacement if the tumor arises from outside this area. Tumors arising in the anterior half of the orbit usually displace the globe to the opposite side and may produce little or no real exophthalmos. The amount of forward displacement of the eyeball can readily be measured with a Hertel's exophthalmometer; 15 - 20 mm. is usually considered normal. Pain is not a common symptom, and congestion of the conjunctiva occurs but rarely, although it may be present in vascular tumors.

Diplopia resulting from proptosis and the resulting immobilization of the eye may not be apparent to the patient because of visual impairment of the affected eye. Visual field defects and loss of visual acuity occur early in tumors invading the optic nerve or optic canal and may occur later in tumors of other parts of the orbit that produce pressure on the optic nerve or globe due to their increased size.

Incidence

The most common orbital tumor is the hemangioma, which is a benign type of growth, is usually well encapsulated, and, though it produces exophthalmos, has little effect on ocular motility. Reese¹ reports 25 in a series of 174 cases of tumor of the orbit. Meningioma and pseudotumor occur next in frequency. The pseudotumor is a little-known tumor and is usually thought to consist of chronic inflammatory tissue. Most of these tumors progress in size to produce exophthalmos and diplopia.

Other tumors occurring frequently are the dermoid cyst, neurofibroma, sarcoma and glioma, and mixed tumors of the lacrimal gland. Of the latter Reese¹ found 9 in his series of 174 cases.

Orbital tumors seldom occur bilaterally, but when they do they are usually of the leukemic infiltration type.

*Genoa, Ohio. Formerly Fellow in Department of Ophthalmology.

Diagnosis

As an aid to diagnosis the following procedure is usually employed: the visual acuity, visual fields, fundus examination, refraction, and muscle balance are first evaluated. By palpation and use of the stethoscope, the tumor is then localized, its consistency is determined, if possible, and bruit and pulsations are ruled out. If a systemic disease is suspected as the underlying cause, a general examination together with complete blood study and basal metabolism test is carried out. A most valuable adjunct to diagnosis is a roentgenogram of the orbit and its surrounding structures. A biopsy, either by aspiration or by operation, may be performed, although this procedure is not often done.

Differential Diagnosis

There are numerous diseases that produce exophthalmos and otherwise simulate orbital tumors.

Exophthalmos caused by high myopia, 25 or more diopters, must be excluded. This condition may be unilateral or bilateral. Early hyperthyroidism with exophthalmos sometimes affects one eye earlier than the other. The method of determining these entities is self evident. Diseases of the accessory sinuses, such as pyocele, mucocele, and hyperostosis of the orbital walls, frequently result in exophthalmos; these may be ruled out by roentgenologic examination. Diseases such as Mikulicz's disease, Hodgkin's disease, xanthomatosis, and syphilis sometimes are confusing and require complete medical evaluation. Pseudotumor, thrombosis of cavernous sinus, and orbital cellulitis cause exophthalmos and usually pain and fever and can be suspected because of the associated symptoms. An aneurysm, either traumatic or due to vascular anomaly, is manifested by a pulsation and bruit over the involved eye.

Treatment

Therapy depends upon the final diagnosis, although treatment is primarily surgical. If all medical causes have been ruled out and differential diagnosis cannot be made, and there is danger to visual acuity, surgical exploration should be done.

The Kronlein² operation has been recommended as the best means of exposing a tumor in the orbit, although this procedure is usually unnecessary; a transconjunctival incision either at the external canthus (with an external canthotomy and resection of the external rectus muscle, if necessary) or an incision in the superior or inferior cul-de-sac is frequently adequate to expose and remove the growth. If the growth appears malignant a specimen of tissue can be removed for microscopic examination. Excision of the growth, enucleation of the eye and the growth, or exenteration can then be done.

Certain tumors, such as the leukemic infiltrations, the xanthomatoses, and pseudotumors, are amenable to x-ray therapy. These are not always diagnosed prior to microscopic examination.

Case Reports

The following 3 cases of rare tumors, a nerve sheath tumor, an adenocarcinoma of the lacrimal gland, and a lymphosarcoma, have been treated in the Department of Ophthalmology during the past year.

The nerve sheath tumor (neurilemmoma) occurs rather infrequently. Reese¹ mentions one in his series of cases, and Rottino and Kelley³ in 1941 reviewed the literature and found but 11 cases reported. They occur at almost any age between 2 and 70 years. Growth is usually progressive.

Case 1. Neurilemmoma. A white woman, aged 50, was admitted to the Department of Ophthalmology on February 11, 1947, complaining of the recent onset of sharp pain in the left side of the face and prominence of the left eye of five years' duration. There was no visual impairment or diplopia. (Figure)

External examination revealed pressure which was forcing the left eye superiorly and temporally and a moderately firm mass just below the eyeball. The anterior-posterior measurement was O.D. 17 mm., O.S. 22 mm.



FIGURE. Case 1. Patient with neurilemmoma of left orbit.

On ophthalmologic examination the visual acuity was O.D. 6/9-2, O.S. 6/12-1, the fundi were normal, and the muscle balance test revealed exophoria of 16 diopters and left hyperphoria of 1 diopter.

Roentgenologic examination revealed some increased density of the left maxillary sinus. Roentgenograms of the teeth gave normal findings. Except for the findings in the left antrum, the ear, nose, and throat examination was negative. Blood and urine were normal. A pre-operative diagnosis of tumor of the left orbit was made. On April 7, 1947, a transconjunctival incision at the external aspect of the left eye with an external canthotomy was made, and a small, well-encapsulated tumor was removed *in toto*.

The gross specimen was an ovoid mass measuring 25 by 15 by 10 mm., encapsulated, and of soft consistency. Tags of fibrous tissue were present on the surface. On section there was a cystic structure 9 mm. in diameter. The tissue was semitranslucent, grayish-yellow to pink in color, and soft and friable in consistency.

On microscopic examination the mass was rather cellular, but there were patches of loose arrangement and slight cellularity. Cells were small, of spindle shape, and arranged in sharply interlacing bundles, and there were frequent areas of nuclear palisading. The tissue bordering the cystic structure was of loose arrangement, and the vessels were moderately numerous, showing thick hyaline walls. There were occasional mitoses. A thin but well defined fibrous capsule was present. A diagnosis of neurilemmoma was made.

The postoperative course was uneventful. When last seen at the Clinic the patient complained of diplopia, and muscle measurement still showed a pronounced error. There was no evidence of recurrence of the tumor.

Lymphosarcoma is also a rare tumor. In Reese's¹ series it occurred in 5 cases, and Dandy⁴ in his monograph on 31 cases of orbital tumors mentions but 1 case (round cell sarcoma), Iles and Short⁵ had no incidence of lymphosarcoma in their series of 14 cases, 3 of which were malignant. This type of tumor usually occurs in later life, the average age being 55 years. It is highly malignant and radiosensitive but unfortunately is only one manifestation of a fatal systemic disease.

Case 2. Lymphosarcoma. A white man, aged 66, was first seen on June 2, 1947, complaining of a growth which had been present on the lateral aspect of the left eyeball for the past five weeks. It caused him to see double on reading or looking to the left. The patient had no pain and did not think his vision was affected. He believed that the growth had increased rapidly in size since he had first noticed it.

External examination revealed a rather firm subconjunctival mass on the temporal side of the left eyeball which seemed to arise inferiorly. No exophthalmos was observed.

On ophthalmologic examination the visual acuity was O.D. 6/6-4, O.S. 6/6-4, and the fundi were normal.

The blood counts were essentially negative except for a slight poikilocytosis; the leukocyte count was 9600. Roentgenologic examination of the chest revealed bilateral infiltration which was thought to be consistent with an old acid-fast lesion.

A preoperative diagnosis of subconjunctival tumor was made, and on June 7, 1947, a friable tumor mass was removed with some difficulty from the inferior temporal aspect of the left eyeball. The tumor tissue appeared to be infiltrating the globe.

The gross specimen consisted of many small portions of tissue, the largest measuring 9 mm. in diameter. Fragments varied from fairly firm to soft texture and were of yellowish-pink color. Several smaller portions were composed of fat and fibrous tissue.

On microscopic examination the fragments were formed principally of striated muscle and there were areas of dense infiltration by small round cells with round nuclei. A moderate number of mitoses were present, and a few eosinophils were found.

A diagnosis of lymphosarcoma was made.

The immediate postoperative course was uneventful, and 250 r was given on each of the ten successive days. Examination for lymphadenopathy elsewhere in the body revealed only two small affected glands in the right groin. A guarded prognosis was given; however, a communication from the patient six months later stated that he was feeling well.

Mixed tumors of the lacrimal gland may occur as adenocarcinoma, and when the epithelial element predominates these are sometimes known as cylindromas. Although this is a carcinomatous tumor it is usually well encapsulated. It usually occurs in persons of middle age or older.

Case 3. Adenocarcinoma of lacrimal gland. A white woman, aged 43, reported to the Department of Ophthalmology on February 24, 1947, giving a history of prominence of the left eye of eight months' duration. No visual impairment or diplopia had been observed. Four months previously she had noticed sharp pains around the left side of the forehead and the left eye.

External examination revealed exophthalmos of the left eye, and a small mass could be palpated under the upper lid. Anterior-posterior measurements were O.D. 19, O.S. 21.

On ophthalmologic examination the visual acuity was O.D. 6/6-3, O.S. 6/6-3. The fundi were essentially normal, and the muscle balance test showed exophoria of 6 diopters for near and $\frac{1}{2}$ diopter for distance. Visual fields were within normal limits.

Ear, nose, and throat examination was negative, and x-ray examination of the sinuses and orbits was normal. Blood, urine, and basal metabolic rate were normal.

A preoperative diagnosis of tumor of the left orbit was made. On March 27, 1947, a firm fibrosed tumor was removed through an incision in the outer aspect of the upper lid.

The gross specimen consisted of a small portion of tissue measuring 15 by 10 by 10 mm., which was irregularly ovoid in shape, white, and of moderately firm, tough consistency. The cut surface presented a somewhat trabeculated appearance, with pinkish-red tissue between the trabeculations.

On microscopic examination the bulk of the tissue was formed by small, dark-staining epithelial cells, variably cuboidal, spindly, polyhedral, or flat. In the more solid areas they resembled basal cells. They were arranged in cords distributed through dense connective tissue as laminated structures with reduplication of lamina. The elements resembled basal cells forming irregular small islands which contained lamina filled or partly filled with pink-staining hyaline material. The pink-staining material occurs as round masses filling or partly filling the lamina. Occasional mitoses were seen. Several nerves were surrounded and infiltrated by the atypical epithelial cells. Along one margin of the section were fragments of normal-appearing lacrimal gland.

A diagnosis of adenocarcinoma, cylindroma in type, of the lacrimal gland was made.

Postoperative x-ray therapy was not deemed useful because of its inefficacy in this type of tissue. The patient is to be observed for recurrence, in which case radical exenteration will be performed.

Summary

The neurilemmoma is a rare type of tumor which may appear at any age and is of progressive growth. The lymphosarcoma is a highly malignant type of tumor which is radiosensitive and may be treated with x-ray alone if the diagnosis is made before operation. Adenocarcinoma of the lacrimal gland is perhaps less rare than the other two types. It is also a malignant growth for which operation may not be completely successful and to which x-ray therapy is ineffective.

The authors gratefully acknowledge the cooperation and assistance of Dr. John B. Hazard of the Department of Pathology.

References

1. Reese, A. B.: Orbital tumors and their surgical treatment. *Am. J. Ophth.* **24**:386 (April); 497 (May) 1941.
2. Elschmig, A.: Operations for tumors of orbit. *Surg., Gynec. & Obst.* **45**:65-73 (July) 1927.
3. Rottino, A., and Kelly, A. S.: Specific nerve sheath tumor of orbit (neurilemmoma, neurinoma); report of case with review of literature. *Arch. Ophth.* **26**:478-488 (Sept.) 1941.
4. Dandy, W. E.: *Orbital Tumors: Results Following the Transcranial Operative Attack.* (New York: G. E. Stechert and Company, 1942).
5. Iles, A. E., and Short, A. R.: Orbital tumors. *Brit. J. Surg.* **31**:147-150 (Oct.) 1943.