A Case of Congenital Orbital Teratoma

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A case of congenital orbital teratoma with marked unilateral proptosis was noted at birth. Patients with congenital orbital teratoma had a uniform clinical picture and were very similar in their morphological pattern, often polycystic and with varied amounts of solid tissue. Six months after birth, the orbital tissues were totally extirpated. Histological examination revealed components from all three germinal layers and no sign of malignancy. Within the first year, the condition of the patient has been satisfactory.

Key words: congenital orbital teratoma, proptosis, three germinal layers, polycystic, solid tissue.

INTRODUCTION

Congenital teratoma of the orbit is a rare condition which manifests itself dramatically by marked unilateral proptosis in a newborn infant. These disfiguring masses can rapidly expand, causing secondary damage to the eye. The tumors consist of fluid and encapsulated cysts mixed with varied amounts of solid tissue. Microscopically they also feature a curious potpourri of well-differentiated tissues derived from two or three germinal layers. The present case is of particular interest because of the rarity and the typification of congenital orbital teratoma.

CASE REPORT

A 6 month old child visited this hospital with a protruding mass in the left orbital region, which had been present since birth. Pregnancy had been full term and normal, and labor and delivery were uneventful. There was no family history that could have had any connection with child's condition.

Clinical Examination

An adult fist-sized mass consisting of nodules was noted in the left orbital area (Fig. 1). The distended palpebral fissure with an everted palpebral conjunctiva, thinning of the eyelid, and a black discoloration beneath the upper eyelid, which was thought to be an eyeball, were observed.

The right eyeball and orbit revealed no abnormality. There were no other abnormalities of evidence of disease.

Radiologic Examination

Simple skull X-ray showed enlargement of the left orbit and bony erosion of the nasal cavity, ethmoid sinus, and maxilla. A rounded soft tissue mass could be seen as a shadow superimposed upon the left orbit (Fig. 3). Computed tomography revealed a large cystic cavity, irregular calcific density, upward displacement of the eyeball, elongation of the optic nerve and extraocular muscles, destruction of the left maxilla and ethmoid, and an invasion of the middle cranial fossa, but no connection with brain parenchyma (Fig. 4).
Surgery

Under the general anesthesia, with the patient in a supine position, 1% lidocaine with 1: 100,000 epinephrine solution was infiltrated beneath the intact skin overlying the tumor.

The skin flaps above and below the tumor were elevated, but tumor invaded the skin was not included in the flap. Meticulous dissection was done between the skin flap and the tumor. Since the tumor had invaded the middle cranial fossa, it was cautiously separated from the dura. The mass and eyeball including the involved maxilla were removed (Fig. 6). Bleeding from the dura was controlled with gelfoam. The opening into the nasal cavity and ethmoid sinus was closed by repairing the remaining mucosa. The exposed dura and the maxilla were covered with the prepared skin flaps. the area which was not covered by the flap was repaired with a split thick skin graft from the left thigh.

Pathologic Examination

The encapsulated tumor measured 9×6×5cm. Bone, cartilage, cyst and hairs were readily identified (Fig. 7).

The eyeball showed the sequelae of compression with atrophy of the stretched optic nerve. The tumor was composed of tissue from all three germinal layers. Endodermal components were represented by several lumina lined by columnar epithelium with mucous secretion (Fig. 8). Mesodermal components were present as connective tissue, vessels, smooth muscle, cartilage, and bone with bone marrow cavities and hematopoiesis (Fig. 9). Ectodermal components were found in the form of skin with hairs and sebaceous glands (Fig. 9). Neuroectodermal tissue, both differentiated and undifferentiated nervous tissue, was also seen. The components of the tumor were all histologically benign.

DISCUSSION

Bonnet, Marchand, Waldeyer, and Krafka have proposed the histogenesis of teratomas. In these, the "organizer" theory (Spemann and Budde) modified by Krafka provides a more
Fig. 4. CT scan demonstrates a large mass occupying the left orbit. Note lucenties suggestive of cystic changes.

Fig. 5. CT scan after operation.

tenable explanation. This theory discusses interference with the normal development of the growth center of the embryo (stated to be the primitive streak in man) allowing the development of a secondary growth center. Migration of this center to different portions of the embryo leads to the occurrence of a teratoma. The rare occurrence of orbital teratomas may be the result of the greater distance between the primordia of the orbit and eye and the primitive streak.

Duke-Elder\(^2\) has classified teratomas of the orbit as follows:

1. A complete fetus implanted in the orbit (or-
Fig. 7. The gross appearance of the orbital tumor.

Fig. 8. This picture reveals intestinal glands lined by a mucin secreting columnar epithelium (hematoxylin-eosin, x100).

Fig. 9. A portion of skin covered with a keratinizing squamous epithelium and a hyaline cartilage with peripheral ossification are seen. The right lower part contains an island of hematopoietic marrow (hematoxylin-eosin, x100).

Fig. 10. This photomicrograph shows a respiratory tract lined by ciliated pseudostratified columnar cells with underlying submucous glands (hematoxylin-eosin, x100).

Fig. 11. Tubuloalveolar lobules like lacrimal glands with mature adipose cells are divided by thin collagen bands (hematoxylin-eosin, x100).

bitopagus parasiticus)
2. A portion of a second fetus in the orbit
3. A tumor consisting of all three germinal layers
4. Tumors containing representatives of two germinal layers only
5. Tumors containing representatives of one layer only.

In most teratomas, the ectodermal and mesodermal components predominate. Endodermal elements are frequently absent or difficult to detect. Jensen proposed that only a tumor derived from all three germinal layers should be called a teratoma. If the tumor is composed of tissue from only two germinal layers, it should be termed teratoid. Tumor with tissue from only one germinal layer in the orbit, i.e. dermoid cyst are choristomas.
A review of the literature showed a female-to-male ratio of 2:1 and a slight preponderance of left sided tumors. Only two cases were bilateral. Two instances of malignant teratoma have been reported. Teratoma is considered malignant when the tissue is embryonal or immature in nature.

The following clinical features are usually noted:

1. Extreme unilateral proptosis with forward or upward displacement of the eye in an otherwise normal full-term newborn infant.
2. Marked stretching of the lids over a tense fluctuant (occasionally solid) mass with elongation of the palpebral fissure.
3. Absence of a demonstrable direct communication between the cavity of the cyst and the intracranial cavity.
4. A normally developed eye which frequently exhibits degenerative changes secondary to displacement by the teratoma.
5. Enlargement of the bony orbit to 2 or 3 times normal size, most often accompanied by displacement of the nose and malar areas from the tumor. In many cases there also were separation of the zygomaticofacial suture and enlargement of the superior and inferior orbital fissures, secondary to local pressure within the orbit.
6. Transillumination of part or all of the orbital mass.
7. Circulatory changes in and about the orbit.

Orbital diagnostic tests include plain radiography of the skull and orbit, hypocycloidal polytomography, computerized tomography, and ultrasonography. Used together, ultrasonography and computerized tomography are correct in up to 98% of all cases of orbital pathology from any cause.

Clinically, it is difficult to distinguish teratomas from other benign and malignant neoplasms. Differential diagnosis includes other congenital anomalies such as dermoid cyst, microphthalmos with cyst, encephalocele, meningoecele. And cystic eye; neoplasms such as neuroblastoma, neurofibroma, hemangioma, and lymphangioma; and other ocular diseases such as congenital glaucoma, retrobulbar hemorrhage, and hemotoma. Radiologically, differential diagnosis includes optic nerve glioma, meningioma, and rhabdomyosarcoma.

The surgical treatment of these lesions almost always has been effective. There are a direct orbital approach through the conjunctiva, intracranial approach, Krüenlein operation, and exenteration. Surgeons now recommend early surgery despite the difficulty in making the diagnosis preoperatively.

Recurrence of continued growth of the incompletely removed teratoma has been reported.

Rapid enlargement of the teratoma after birth is characteristic. The solid portions increase in size, and the amount of fluid increased. When untreated for a period of months, the tumor may become enormous. With the expansion of the mass, the eye becomes exposed, compressed and eventually perforated.

Despite the alarming appearance, it has been noted repeatedly that the infant remains quite healthy and is undisturbed by the tumor so long as secondary infection does not occur.

In our case of congenital orbital teratoma, histological examination revealed components from all three germinal layers and no sign of malignancy. Exenteration was performed at 6 months of age because the teratoma was huge, and the optic nerve was stretched and atrophic. The postoperative period was uneventful. One year later the child was well (Fig. 2).

REFERENCES