INTRAOCULAR MALIGNANT TERATOID MEDULLOEPITHELIOMA

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ABSTRACT

Malignant teratoid medulloepithelioma is an uncommon unilateral intraocular tumor, occurring typically in children. This report concerns a 4-year-old boy, who presented with loss of vision, pain and proptosis of the left eye and showed mass in between eyelids. Histopathologically, the tumor was composed of pseudostratified primitive-appearing epithelium dispersed in cords, strands, tubules & glands, which were separated by a cystic spaces, filled with pale eosinophilic material. Foci of glial tissue, cartilage, bone, skeletal muscle and fatty tissue were recognized. In addition, scleral extension of tumor, Homer-Wright like and Flexner-Wintersteiner like rosettes and foci of necrosis were also present. The differentiation from retinoblastoma was discussed.

Key Words: *Malignant medulloepithelioma, eyeball, heteroplasia.*

INTRODUCTION

Intraocular teratoid medulloepitheliomas are very rare embryonal tumors that usually originate from the ciliary epithelium¹⁻⁷ and rarely from the iris,⁸ retina^{9,15} or optic nerve.¹⁰ Medulloepithelioma is an unilateral ocular tumor,⁸ typically found in children. It accounts for almost all the tumors in the congenital group, but they are much rarer than

retinoblastomas, which are believed to be congenital tumors of the retina. Medulloepitheliomas can be classified according to the WHO as medulloepithelioma or teratoid medulloepithelioma, benign or malignant. The teratoid variant shows heteroplasia with areas of hyaline cartilage, rhabdomyoblasts, undifferentiated mesenchymal tissue or neuroglial tissue. Clinical presentations include poor vision

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& pain; white pupil,¹¹ a mass in the iris, anterior chamber or ciliary body, glaucoma,¹² cataract,¹³ lens coloboma,¹³ iris neovascularization,¹³ ectopia lentis.¹⁴ proptosis etc.

This case intraocular malignant teratoid medulloepithelioma is herewith reported for its rarity.

CASE HISTORY

The patient was a four year old boy, who presented with loss of vision, pain and proptosis of the left eye since last 2 yrs. Later on, proptosis had been progressed and a mass appeared inbetween eyelids. This mass slowly enlarged in size and presented as fungating mass since last 2 weeks.

On examination left eye showed fungating mass, with superficial necrosis. Right eye was apparently normal. No family member or relatives have been diagnosed to have eye tumors in the past. Systemic examination didn't reveal any abnormalities. No regional lymphadenopathies were noticed. Routine blood examination revealed Hb-10.7gm/dl; WBC-9,400/cmm with 55% neutrophils, 41% lymphocytes, 3% monocytes and 1% eosinophils; ESR-17mm/ 1st hour. Ultrasound scanning of the abdomen (liver, gall bladder, spleen, pancreas) were normal. The case was clinically diagnosed as retino-blastoma. As there was a large fungating eyeball mass, protruding inbetween eyelids and vision on this eye was completely absent; the exenteration was carried out.

HISTOPATHOLOGICAL FINDINGS

Gross: Exenteration specimen consisted of eyelids and an eyeball, replaced by a mass, which extended beyond the limits of eyelids. Mass was grey-white & soft, measured 5cm x 5cm x 3cm. It had irregular border and was attached to the wall of eyeball in

the region of ciliary body. Cut surface showed areas of haemorrhage & necrosis.

Microscopic: Tumor was composed of pseudostratified primitive-appearing epithelium dispersed in cords, strands, tubules & glands (fig.1).

Fig. 1

Fig. 1, Showing medullary epithelium in tubules, glands & cords.

These structures were separated by a cystic spaces, filled with pale eosinophilic material (fig.2).The

Fig. 2

Fig. 2, Showing cystic spaces, filled with eosinophilic material.

cells were pleomorphic, columnar shaped with few mitoses. In addition, there were foci of glial tissue (fig.3), cartilage (fig.4), bone (fig.5) skeletal muscle and fatty tissue. There was extension of tumor to the sclera. A few Homer-Wright like and Flexner-Wintersteiner like rosettes (fig.6) were also present. There were areas of focal necrosis. So, histopathological diagnosis of malignant teratoid medulloepithelioma was made.

Fig. 3

Fig. 3, Showing glial tissue as heteroplastic element.

Fig. 6

Fig. 6, Showing Homer - Wright like & Flexner - Wintersteiner like rosettes.

Fig. 4

Fig. 4, Showing lobules of cartilage as heteroplastic element.

Fig. 5

Fig. 5, Showing bony tissue as heteroplastic element.

DISCUSSION

Verhoeff⁽⁹⁾ in 1904 gave the first detailed histological description of a medulloepithelium, but he named the tumor a "teratoneuroma" despite the fact that its histology was not teratomatous. In 1908, Fuchs¹⁶ coined the term "diktyoma" to describe netlike pattern of interlacing ribbons of poorly differentiated neuroepithelial cells. The term "medulloepithelioma" was probably first used for this tumor by Grinker⁹ in 1931.

Medulloepitheliomas are neoplasms containing multilayered sheets & cords of poorly differentiated neuroepithelial cells. These sheets & cords often fold back upon themselves so that structures of varying shapes & sizes are formed. The proliferating medullary epithelium may form intricately anastomosing cords & sheets separated by cystic spaces (containing hyaluronic acid) producing net-like appearance.⁴ Homer-Wright and Flexner-Wintersteiner rosettes may also be observed. Our case was typical and we found all these histological features. The heteroplasia is observed in teratoid medulloepitheliomas. The heteroplastic elements include hyaline cartilage,^{4,17} rhabdomyoblasts,^{4,17} undifferentiated mesenchymal cells,¹⁷ neural tissue⁴ etc. The common heteroplastic element is hyaline cartilage. In our case, the

observed heteroplastic elements were hyaline cartilage, glial tissue, bony tissue, fatty tissue and skeletal muscle. The medulloepithelioma is considered as malignant⁹ if it contains areas composed of poorly differentiated neuroblastic cells with greater pleomorphism & mitotic figures and/or sarcomatous areas resembling rhabdomyosarcoma, chondrosarcoma or embryonal sarcoma. Invasion of uvea, cornea or sclera with or without extraocular extension is also criterion for malignancy. In our case, we found poorly differentiated neuroblastic cells with scleral extension, few Homer- Wright like and Flexner-Wintersteiner like rosettes. However, sarcomatous areas were not observed. Electron microscopically¹⁸ the lumens of the rosettes contain slender microvilli, bordered by terminal bar complexes. Zonula adherens and zonula occludens type junctions are also evident. Electron microscopic study was not done in our case. Immunohistochemically¹⁹ the neuroepithelial tumor cells were positive for neuron-specific enolase, vimentin and often for S-100 protein; and the neuroblastic cells for neuron-specific enolase and synaptophysin. Immunohistochemical study was not done in our case.

Clinically poor vision, pain, mass in iris or ciliary body, proptosis and leukocoria are common features. Our case presented with loss of vision, pain and proptosis of eyeball. The treatment of intraocular medulloepithelioma is surgery i.e. local excision, enucleation or exenteration.⁴ Radiation appears to have no effect. In our case, exenteration was carried out and patient was lost to follow-up.

Distinction from the common ocular tumor of childhood, the retinoblastoma may be sometimes difficult clinically as well as histopathologically. Medulloepithelioma differs from the retinoblastoma by a later age of onset, anterior development, a frequent cystic structure and

presence of heteroplastic elements.^{20,21}

This case report emphasizes the fact that intraocular medulloepithelioma should be considered as a differential diagnosis for an eyeball mass.

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