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Case Note

Malignant teratoid medulloepithelioma in eye

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ABSTRACT
Malignant teratoid medulloepithelioma is an extremely rare tumor occurring in children younger than 5 years of age, arising from ciliary body epithelium or iris but few arise from optic nerve and retina. This report concerns a 5 years old boy who presented with pain, redness and protrusion of right eye. Histopathologically, the tumor was composed of epithelial and sarcomatoid component. The pseudostratified primitive appearing epithelial cells were arranged mainly in diffuse pattern, nests cords and tubules. At places, pseudo rosette and true rosette were seen. Mitoses were frequent consisting of 7-10/ HPF. The sarcomatoid component consisting of spindle shaped cells arranged in interlacing bundle were also seen. Mitoses counted 5-7/HPF. Massive areas of necrosis and hemorrhage along with calcification, focal area of mature cartilage were present. Vascular and optic nerve invasions were seen. This case of malignant teratoid medulloepithelioma is the second case diagnosed in TU Teaching Hospital within the period of 10 years and reported because of its rarity. The differentiations from other tumors of the orbit such as small cell tumor were discussed.

Key words: Malignant teratoid medulloepithelioma, heteroplasia, enucleation.

Malignant teratoid Medulloepithelioma in eye is a very rare congenital[1] tumor of embryonal neuroepithelial origin. The majority of such tumors arise in the ciliary body epithelium [2-5] and few from the iris[6], retina [7,8] and optic nerve[9]. They are unilateral[6] tumors, very aggressive with similar rates of occurrence in each eye. There is no sex & race predilection and hereditary link either. The median age of clinical manifestation of medulloepithelioma is 3.8 years with the range of 6 months - 41 years.
Medulloepithelioma can be classified (WHO) as teratoid or non-teratoid Medulloepithelioma, benign or malignant. The teratoid variant arise due to the pleuripotential nature of medullary epithelium and show heteroplasia with areas of hyaline cartilage, rhabdomyoblast, undifferentiated mesenchymal tissue or neuroglial tissue, the last being the most common.

Generally, the patient presents with pain and poor vision. Clinical signs are leukocoria, visible mass in the iris, anterior chamber or ciliary body, proptosis or pupillary dysfunction glaucoma etc. This case of Malignant Teratoid Medulloepithelioma in eye is the second case diagnosed in Tribhuvan University Teaching Hospital within the period of 10 years

Case History

The patient is 5 years old Muslim boy who presented with whitish pupil at the age of 2 months and was diagnosed as unilateral cataract.
Three months back, the patient was presented with the complaints of painful right eye with redness and protrusion for 20 days.
On examination, right eye showed protrusion with corneal necrosis. Left eye was apparently normal. Systemic examination did not reveal any abnormalities. No regional lymphadenopathies were noticed. There was no significant family history. Routine blood examination revealed Hb-12.4gm/dl, WBC-6500/cmm with 50% Neutrophil, 40% Lymphocytes, 2% Monocytes, 8% Eosinophils, Normal CXR finding. The case was clinically diagnosed as Retinoblastoma with orbital extension and enucleation was carried out.

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Histopathological finding

Gross

Enucleation specimen consists of eyeball along with eyelids and adherent fatty tissue measuring 5x4x2 cm. Eyeball is grayish brown in color. Rest of the portion is organized into a solid mass measuring 2.5x2 cm. Optic nerve measures 0.9x0.3 cm. Cut section shows whitish area with area of hemorrhage and necrosis. Gritty sensation felt on cutting at places.

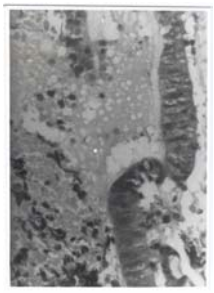

Fig. 1

Tissue section showing primitive appearing epithelium arranged in tubules magnification x400, H/E stain

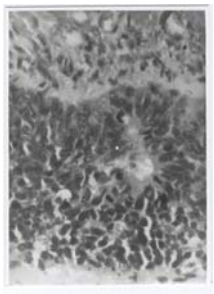

Fig. 2

Tissue section showing tumour cells arranged in rosette with numerous mitosis. magnification x400, H/E stain.

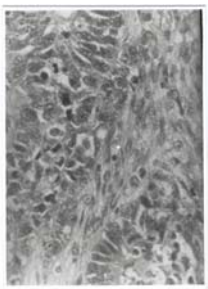

Fig. 3

Tissue section showing sarcomatoid component with mitosis. magnification x400, H/E stain.

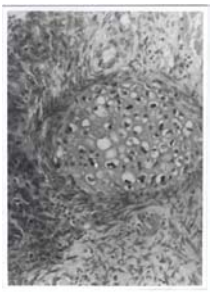

Fig. 4

Tissue section showing mature cartilage in the center. magnification x400, H/E stain.

Microscopic

Tumor was composed of epithelial and sarcomatoid component. The pseudostratified primitive appearing epithelial cells were arranged mainly in diffuse pattern, nests, cords, tubules along with true and false rosettes. The cells are round, elongated to polygonal in shape, intermediate to large in size with abundant eosinophilic cytoplasm. Nucleus is large and hyperchromatic and frequent mitoses (7-10/HPF).

The sarcomatoid components, consisting of spindle shaped cells were arranged in interlacing bundles. These tumor cells had moderate amount of cytoplasm and spindle shaped nuclei with blunt end and coarsely clumped chromatin. Mitoses were 5-7/HPF.

Massive area of necrosis, hemorrhages along with calcification, focal area of mature cartilage were present. At the periphery stratified squamous lining with fibrocollagenous tissue was seen. Normal fibrous tissue was present in between the tumor cells. Vascular and optic nerve invasions were seen. So, histopathological diagnosis of malignant Teratoid Medulloepithelioma infiltrating into optic nerve was made.

Discussion

Medulloepitheliomas are very rare congenital tumor of embryonal neuroepithelial origin. It was first described in 1904 by Verhoff[7] as a teratoneuroma and later renamed as Medulloepithelioma by Grinker[7] in 1931. Medulloepithelioma appear as multilayered sheets and cords of poorly differentiated primitive neuroepithelial cells. Homer Wright and Flexner-Wintersteiner rosettes are also observed. In our case all these histological features are seen. These epithelial like components can be shown immunohistochemically [10,11] and stained positive for NSE, S100, GFAP & EMA on the luminal border.

Heterotropic tissues like hyaline cartilage[3], rhabdomyoblast[3], undifferentiated mesenchymal tissue[3] or neuroglial tissue[3] are observed in teratoid medulloepithelioma. In our case mature cartilage and fibrous tissues are seen. Neuroglial component will be stained positive for Synaptophysin, GFAP and S100 and rhabdomyoblastic component will be stained positive for Desmin and MSA. Immunohistochemical stains were not done in our case.

Medulloepithelioma are considered as malignant[7] if it show 1) areas of poorly differentiated neuroblastic cells resembling neuroblastoma, 2) increased pleomorphism or mitotic activity, 3) sarcomatous areas, 4)

invasion of uvea, cornea, or sclera with or without extraocular extention.

In our case pseudostratified primitive appearing epithelial cells with increased pleomorphism & high mitotic count around 7-10/HPF, sarcomatous areas and vascular and optic nerve invasion with tumor cells are seen.

Generally the patient presents with pain and/or poor vision and clinical signs are leukocoria, visible mass in the iris, anterior chamber or ciliary body, proptosis etc. Our case presented with pain redness and protrusion of the eye. Treatment of choice of malignant teratoid medulloepithelioma is surgical enucleation[3] which was done in our case. Patient was discharged and lost to follow up.

The prognosis for children with classic ciliary medulloepithelioma is fairly good however for tumors that present in rare sites like the retina or optic nerve, prognosis is not so good. They tend to more readily spread intracranially which is the most common cause of death in any medulloepithelioma. This neoplasm is reported to be locally aggressive and capable of recurrences, but distant metastases are very rare.
Whenever encountered with the tumors of the orbit, small cell tumor such as retinoblastoma, neuroblastoma, malignant melanoma should be considered as differential diagnosis. Retinoblastoma can be diagnosed with the features of small round cells with hyperchromatic nuclei arranged in trabeculae and nests with Wintersteiner rosettes and Neuroblastoma with bipolar like cell elements with hemorrhage, calcification and Homer Wright's rosettes. But in our case, epithelial cell with pleomorphism and high mitotic count, sarcomatoid component with heterotropic element like cartilage are present. Malignant melanoma can be differentiated with the features of prominent melanin pigment, nuclear grooves, folds and pseudoinclusion which is absent in our case.

Sometimes, it may be difficult to differentiate even histopathologically for which clinical information and microscopical features including heteroplastic tissue is very important.

This case report emphasize the fact that Malignant teratoid medulloepithelioma should be considered as differential diagnosis for any eye ball mass.

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