

Letters to the Editor

Commentary on Immunohistochemical Diagnosis of Rhabdomyosarcoma

Dear Editor:

Hehua Ye and colleagues reported an interesting and rare case of primary conjunctival rhabdomyosarcoma (RMS) successfully treated with surgery and chemotherapy in Iranian journal of ophthalmology 2011;23(4):65-68. There are some considerable comments that I would like to point out.

Orbital RMS is the most common malignant mesenchymal neoplasm of the childhood and may also rarely present as a primary subconjunctival mass. However on histopathological examination, the diagnosis of RMS is the most suspicion in this case, nevertheless a suitable panel of immunohistochemical studies was needed to definite diagnosis of RMS and rule out other lesion which should be considered in differential diagnosis of mesenchymal tumor of the conjunctiva in childhood, with better prognosis such as fibroblastic and myofibroblastic lesions.^{1,2} In the reported case, primitive tumoral cells were spindle shape in routine H&E stain and only immunoreactive for vimentin, desmin and nonspecific actinin in IHC staining, these findings were not specific evidence of RMS. There are large panel of relatively specific immunohistochemical markers with different specificity and sensitivity including: myogenin, sarcomeric actin, myosin, myoglobin, etc for diagnosis of RMS but for routine investigation of RMS, myogenin, sarcomeric actin and desmin are chosen.³

Actin is an extremely useful marker for the identification of smooth muscle cells, and myofibroblast.

Various isoforms of this protein exist, including those which are specific for smooth muscle and striated muscle. Antibodies specific for striated muscle (sarcomeric) actin are used as markers for RMS.³

Desmin (an intermediate filament) is found in both cells of smooth and striated muscle and in a lesser amount also in myofibroblast. It is particularly abundant in parenchymal smooth muscle.³

Vimentin is characteristic of cells of mesenchymal nature, such as endothelial cells, fibroblasts and vascular smooth muscle cells sometimes also expressed in tumors of epithelial or neural nature, so it lacks specificity as striated muscle marker.³

Myogenin is a myogenic nuclear protein and one of the top choices for identification of skeletal muscle differentiation in tumors and has a high degree of specificity.³

Myoglobin is apparently specific for striated (skeletal and myocardial) muscle, therefore, of utility for identification of RMS and other tumors exhibiting skeletal muscle differentiation.³

Spindle cell rhabdomyosarcoma is a rare subtype of rhabdomyosarcoma mostly occurring in children and adolescents which is categorized as a variant of embryonal RMS in the 2002 WHO classification. It is placed in the superior prognosis category of the international prognostic classification of pediatric RMS.

In contrast to other types of RMS, the 5-year survival is over 95%.⁴

In addition a complete and suitable panel of immunohistochemical studies for exact diagnosis of RMS, this rare variant of rhabdomyosarcoma (spindle cell type) was desirable considered on discussion of reported case.

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