Case Report

A Revisit to Foetal "Rhabdomyoma": An Electronmicroscopic and Immunologic Evaluation

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The fetal "rhabdomyoma" is a rare tumor occurring most commonly in the scalp of young children\(^1\). The tumor closely mimics embryonal rhabdomyosarcoma by light microscopy, however, it lacks frequent mitoses and pursues a benign course. We examined a case of classical “Fetal Rhabdomyoma”. In addition to the routine histopathology, immunoperoxidase and electronmicroscopic studies were carried out. Our findings negate the origin of this tumor from skeletal muscle and support its origin from fibroblast. The presence of markedly dilated endoplasmic reticulum in fibroblasts gave a false impression of cross striation on light microscopy. We propose that the name of this lesion may be changed from Fetal Rhabdomyoma to Fetal Fibroma.

Report of A Case

A 5 month old and 6.5 Kg. male Saudi infant, product of full term normal spontaneous vaginal delivery was presented with a swelling over the left temporal region. The mother noted the swelling at birth however it increased in the last two months prior to coming to the hospital. On examination the child was alert. His head circumference was 42 centimeter. The swelling was 4 cm in diameter, subcutaneous, slightly moveable, non-pulsating and non-tender.

The laboratory findings included WBC: 5.8 X 10^3/mm\(^3\), RBC: 4.64 X 10^6/mm\(^3\), hemoglobin: 11.3 g/dl, HCT: 3.3 %, MCV: 71.7 fl, MCH: 24.3.

The tumor was excised under general anesthesia. The tumor could be easily separated from the scalp fascia and from the underlying bone. However it appeared to be attached to the periosteum. A small amount of the periosteum was removed along with the specimen. The underlying bone appeared to be somewhat hyperemic and there were a few small communicating channels through the cranial bones.

Surgical Pathology Examination

The tumor was 4 X 3 X 2 cm, well-circumscribed, tan pink, soft, slightly lobulated, somewhat gelatinous and avascular (Figure 1). The cut sections revealed glistening and shining surfaces. On light microscopy the tumor was quite cellular, being composed of mostly undifferentiated spindle shaped cells with scant cytoplasm (Figure 2). However occasional cells were racket and tadpole shaped. Their cytoplasm was abundant and eosinophilic. There was suggestion of cross striations (Figure 3). The stroma was loose and edematous. At places however the cells were arranged in compact areas. The nuclei were large with dense chromatin. Mitoses were extremely rare. No abnormal mitosis was seen. The tumor cells were faintly PAS positive. The trichrome stains showed brownish red nuclear and cytoplasmic staining.

The Immnoperoxidase staining with the antimyoglobin stains were completely negative.
tumor was positive for vimentin and negative for cytokeratin and protein S 100.

Fig. 1: Gross appearance of the tumor.

Fig. 2: Light microscopy of the tumor showing undifferentiated spindle shape cells. The cells are forming compact layer as seen in the embryonal rhabdomyosarcoma. (H&E X 250)

Fig. 3: The racket or Tad pole shaped tumor cells. Note the cross striations (Arrow). (H&E. X 400)

Fig. 4: The ultrastructure of the tumor cells. The nuclei are eccentric. The cytoplasm contains prominent dilated rough endoplasmic reticulum. (X 5000)

The electron microscopy revealed the tumor cells to contain eccentric nucleus with peripheral condensation of the chromatin. The nucleoli were not prominent. The cytoplasm was chock-full of prominent dilated granular endoplasmic reticulum (Figure 4). Occasional mitochondria were also present. There was no evidence of thick (actin) and thin (myosin) filaments.

Discussion
The rhabdomyoma is a rare tumor. It mostly occurs in the heart. A distinct entity of rhabdomyoma is seen in the very young children that is termed as “Fetal Rhabdomyoma” They occur almost exclusively in very young male children and infants. Occasionally the babies are born with these lesions. The post auricular region of the scalp is the most characteristic site. The lesion is usually well-circumscribed gray white to tan pink, and soft in consistency. The underlying bones may be slightly hyperemic but not infiltrated by the tumor. Microscopically the tumor closely mimics the embryonal rhabdomyosarcoma because of spindle shaped, undifferentiated cells forming compact layers. However the cells are less bizarre and show neither increased nor abnormal mitoses. Under the light microscope faint cross striations can be seen in some cells.
The present case fulfills all the criteria described for the fetal rhabdomyoma. The age, sex, location and microscopic appearance were very typical. However the immunoperoxidase and electronmicroscopic studies failed to confirm the rhabdoid myogenous origin of the tumor. On EM no myofilaments were identified. On the other hand the most prominent ultrastructure feature was the presence of a large number of rough endoplasmic reticulum that constituted the bulk of the cytoplasm. It seems that these prominent profiles of the dilated ER were giving the appearance of the "cross striations" at the light microscopy level.

The fetal rhabdomyomas are considered by some, hamartoma, rather than a true neoplasm. The argument put forward is that in some cases these may persist for years without increasing in size or causing any discomfort to the patient. Several other pathologic lesions are known to be frequently mistaken for the rhabdomyomas. These include oncocytomas and decidual polyp of the vagina. Invariably such lesions contain large cells with eccentric nuclei and deeply eosinophilic cytoplasm with occasional suggestions of cross striations thus closely resembling the differentiating skeletal muscle cells. However the cross striations at the light microscopy level may be deceptive at times as was the case here. Active fibroblasts and other cells with prominent profiles of dilated RER reticulum may appear to contain faint cross striations at the light microscopy level. We therefore stress the need for confirmation of rhabdomyoid origin by immunohistochemistry and ultrastructural investigations.

In summary, a case of a typical "foetal rhabdomyoma” is presented here. The tumor appears to be of fibroblastic rather than rhabdomyomal origin. There is need to carry out ultrastructural and immunoperoxidase studies on all these cases. If rhabdoid origin is not seen in most of these cases then the tumor may be regarded of the origin for fibroblasts and term may be changed from Foetal Rhabdomyoma to Foetal Fibroma or fetal fibromatosis.

References