

SECTION EDITOR: ENID GILBERT-BARNES, MD

Pathological Case of the Month

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A 2-YEAR-OLD white boy had an 8-mm nodular lesion on his tongue since birth. The nodule was completely resected. No signs of recurrence were noted at 7-year follow-up. The specimen consisted of 2 mucous-covered fragments measuring 1 and 2 cm in diameter. Microscopically, there was a moderately well-circumscribed, 7 × 6 × 4-mm nonencapsulated, submucous nodular lesion (**Figure 1**). The central portion of the nodule was composed of thin, elongated cells with eosinophilic cytoplasm arranged haphazardly in slender fascicles or isolated. Small oval-to spindle-shaped undifferentiated cells with indistinct cytoplasm were interspersed among them (**Figure 2**). A decrease in the number of the undifferentiated cells was noted toward the periphery; the elongated cells became larger and disclosed a greater degree of cytoplasmic differentiation (**Figure 3**). Both types of cells showed bland nuclei with

no mitoses and inconspicuous nucleoli. A discrete amount of myxomatous stroma were noted, especially in the central portion of the lesion. There were no areas of necrosis or inflammation. Immunohistochemical stains were obtained (**Figure 4** and **Figure 5**).

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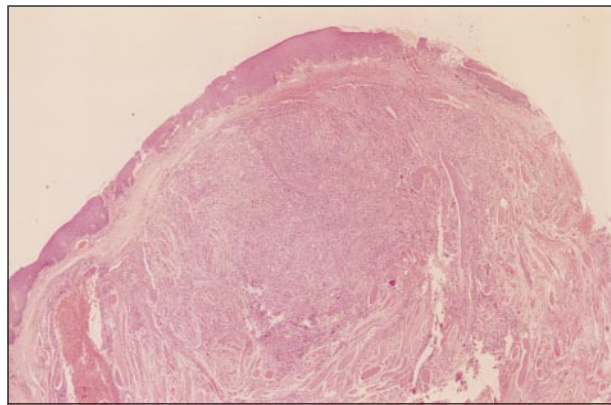


Figure 1.

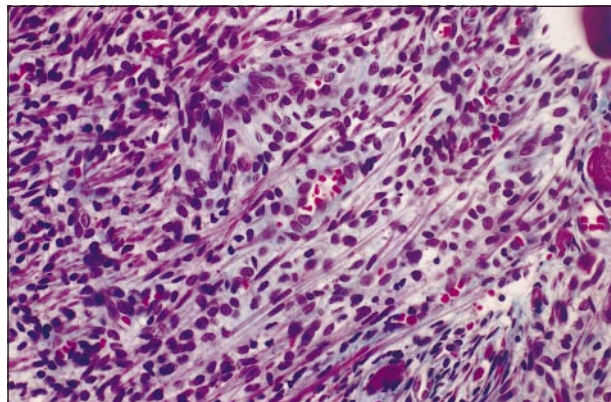


Figure 2.

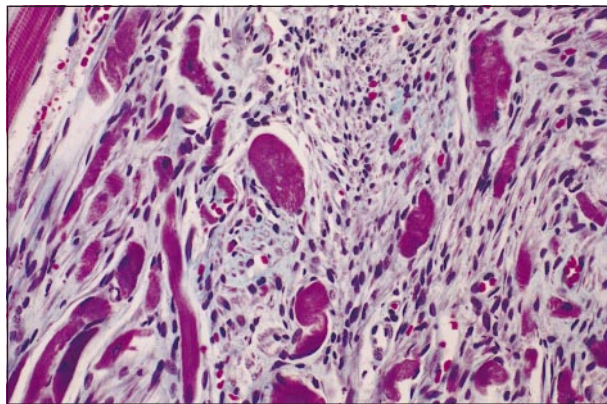


Figure 3.

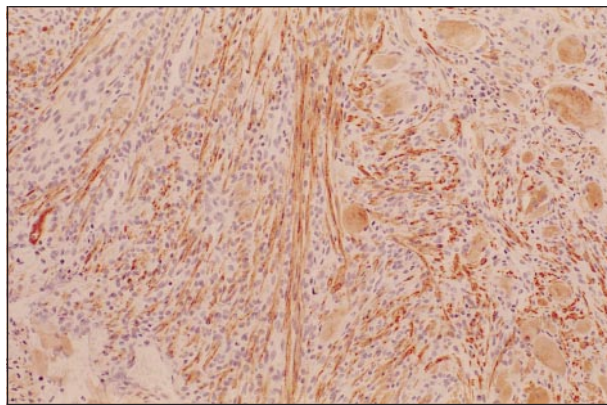


Figure 4.

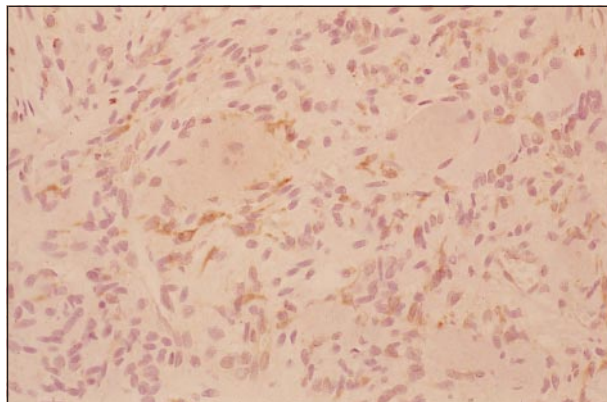


Figure 5.

Diagnosis and Discussion

Fetal Rhabdomyoma of the Tongue (Myxoid Type)

Figure 1. Note the nonencapsulated nodule between muscle fibers of the tongue preserving the mucosa (hematoxylin-eosin, original magnification $\times 20$).

Figure 2. The elongated purple-red cells correspond to immature skeletal muscle fibers without cross striations. The small cells with scant cytoplasm correspond to undifferentiated mesenchymal cells. Both types show benign nuclei without atypia or mitosis (Masson trichrome, original magnification $\times 400$).

Figure 3. Skeletal muscle fibers are more differentiated toward the edge with prominent cytoplasm and cross striations. There is a decrease in the number of mesenchymal cells. Note normal muscle fibers at the left (Masson trichrome, original magnification $\times 200$).

Figure 4. Myofibers show intense intracytoplasmic immunorepression for muscular markers, such as actin (peroxidase, original magnification $\times 100$).

Figure 5. Cytoplasm of the undifferentiated mesenchymal cells stain faintly for glial fibrillary acidic protein (peroxidase, original magnification $\times 400$).

Rhabdomyoma is a benign neoplasm of striated muscle tissue. It is seen less frequently than its malignant counterpart, rhabdomyosarcoma, in contrast to other soft tissue tumors for which benign neoplasms are seen more frequently than malignant counterparts.^{1,2} Rhabdomyomas can be classified topographically as cardiac and extracardiac types. There are 4 different types of extracardiac rhabdomyoma: adult, fetal (least common³), genital, and rhabdomyomatous mesenchymal hamartoma.^{2,4,5} Fetal rhabdomyoma (FR) is not an early stage in the development of adult rhabdomyoma,² but it may develop into a rhabdomyosarcoma.^{1,2} It is a rare manifestation of nevoid basal cell carcinoma syndrome, suggesting that it may be a hamartomatous process.^{2,6} It shows 2 recognized histological patterns: the myxoid or classic type (affecting infants during the first year of life and localizing in the subcutaneous tissue and submucosa of the head and neck region, especially the preauricular and postauricular zone) and the intermediate, cellular, or juvenile type (affecting adolescents and adults and involving mucosal sites or soft tissues of the face).^{2,3,5,7} It has also been reported in the upper extremity, urethra, chest wall, abdominal wall, axilla, thigh, stomach,³ retroperitoneum,^{3,6} and anus.⁷

Dehner et al⁸ defined immature or fetal-appearing rhabdomyomatous tumors as FR, characterized by the presence of immature skeletal muscle fibers and primitive small, round to oval mesenchymal cells in a myxoid background stroma. This pattern was interpreted as the myxoid variant of FR. Crotty et al⁴ reported 2 tumors in children with more differentiation and uniformity than myxoid FR. They proposed the term *juvenile rhabdomyoma*. Later, Kapadia et al⁵ described 16 tumors that displayed more advanced rhabdomyoblastic maturation than myxoid FR. They called the tumors FR with *intermediate* differentiation rather than proposing a new category.

Myxoid FR is composed of primitive oval or spindle-shaped cells, interspersed muscle fibers, and a myxoid background. However, the myxomatous stroma is not necessary for the diagnosis of myxoid FR.⁹ The muscle fibers resemble the myotubular stage of muscle development with thin, elon-

gated, eosinophilic cytoplasm, bland nuclei, and differentiation toward the periphery. The intermediate type is identified by numerous differentiated muscle fibers, inconspicuous primitive mesenchymal cells, and sparse myxoid stroma. The muscle cells show abundant eosinophilic cytoplasm with patent cross striations. There are transitional forms between the 2 types.^{2,7,9} Fetal rhabdomyoma expresses desmin, muscle-specific actin, and myoglobin.⁵ Primitive mesenchymal cells variably express S100 protein, glial fibrillary acidic protein,⁵ and vimentin.¹

The main differential diagnosis is the embryonal and the spindle-cell variant of rhabdomyosarcoma.^{1,3,5,7} Fetal rhabdomyomas are superficially located, well-circumscribed, generally without infiltrating margins.^{1,5} Most cases do not contain mitotic figures, and necrosis is generally absent.^{1,3,5,7} The absence of marked nuclear atypia is the most important criterion to distinguish FR from rhabdomyosarcoma.⁵ Other diagnoses such as rhabdomyomatous mesenchymal hamartoma of the skin, benign Triton tumor, infantile fibromatosis (desmoid type), and traumatic granuloma of the oral cavity should be discarded.^{5-7,9} The treatment is local excision. Recurrence is extraordinarily rare, but increases notably if the exeresis is incomplete or infiltrating margins exist.¹

We report, to our knowledge, the fourth well-documented case of FR affecting the tongue.^{1,5,10} We classified it as myxoid FR because its cellularity resembles this type, although there was scarce myxomatous stroma. It is important to be aware of FR because it could be mistaken for malignant tumors. The diagnosis of nevoid basal cell carcinoma syndrome must be in mind especially if the localization is unusual or multicentric.

Accepted for publication August 19, 1999.

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