Fetal intermediate rhabdomyoma of the lip: case report

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Fetal rhabdomyoma is a rare benign neoplasm of skeletal muscle that must be distinguished histologically from various malignant lesions, including rhabdomyosarcoma. They have recently been subdivided into two histological variants myxoid and cellular fetal rhabdomyomas. This article describes a case of an 8 month infant with a fetal rhabdomyoma in the upper lip. Illustrate the histological features and discusses the means of distinguishing these lesions from various other conditions with which they may be confused.
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INTRODUCTION

Rhabdomyomas are rare benign tumors. They do not exceed 2% of all striated muscle tumors[1,2] and are considerably less common than rhabomyosarcomas.[3] They are classified as cardiac and extracardiac. Among the extracardiac rhabdomyomas, four clinically and morphologically different types can be distinguished: 1) adult type, 2) fetal type, 3) genital type, and 4) rhabdomyomatous mesenchymal hamartoma.[4,5]

CASE REPORT

Male of 8 months of age that was referred to the Oral and Maxillofacial Department of the Central Hospital "Dr. Ignacio Morones Prieto" in the city of San Luis Potosí, Mexico with a lesion, bounded in left region of superior lip, present since birth, which has stayed stable in size. It was primarily seen at the second month of age with the probable diagnosis of an inclusion cyst vs leiomyoma and the staff decided to maintain the mass under observation and the surgery was programmed when the patient reached eight months.

Under general anesthesia the surgical procedure started with an incision in the vestibular mucosa of the upper lip. Dissecting until finding a solid mass on the muscular plane, not encapsulated and with a size of about 2.5 cm of diameter (Figure 1). After the local resection was achieved, a reconstruction of the upper lip was also performed (Figures 2, 3).

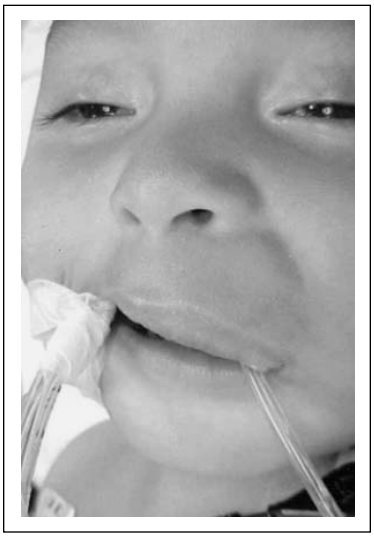

Figure 1. Clinical aspect of the patient where there is a evident mass in the upper lip prior to surgery.

The specimen of approximately 2.5cm of diameter was sent for histopathological study. The excised tissue was fixated in 10% buffered formalin and embedded in paraffin. Tissue sections were stained with hematoxylin and eosin and acid-Schiff. (Figure 4)

Microscopically, the mass showed a proliferation of rhabdomyocytes in a background of dense connective tissue and extensive vascularity. The rhabdomyocytes were round and fusiform and had a eccentrically placed, oval nuclei without significant pleomorphism. Cross-striations were present within the cytoplasm on hematoxylin-eosin stained sections and were highlighted by trichrome stain. No significant mitotic activity was identified. A diagnosis of Fetal Rhabdomyoma.(Figures 5, 6) was made.

DISCUSSION

Rhabdomyoma is a benign neoplasm that contains striated muscle fibers as well as small primitive cells and round or oval mesenchimal cells. This tumor is less frequently seen than rhabdomyosarcomas.[3]

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Figure 2. Two weeks postoperative view of the patient.

Figure 3. Six months postoperative view of the patient.

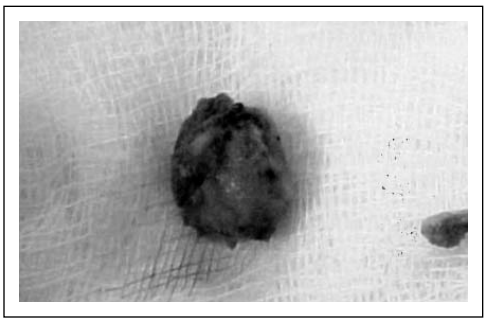

Figure 4. Macroscopic view of the excised lesion of aprox. 2.5 cm. of diameter.

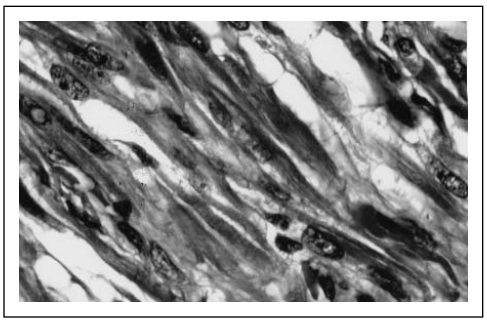

Figure 5. Hematoxilin-eosin stained microscopic view which shows polygonal and rounded cells with a clear stroma.

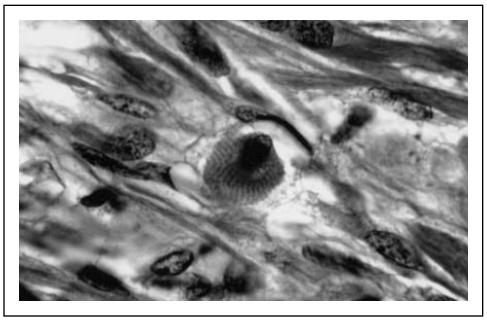

Figure 6. Histological view with oversized round cells with dense round nuclei.

Fetal rhabdomyoma can be classic, myxoid, cellular, intermediate or juvenile. and are very rare,[6] most of the time they can be seen on head and neck, around the ears and sometimes they can be part of the nevoid basal cell carcinoma syndrome.[7,5] The principal problem with this variant is the confusion that can arise with the embrionary rhabdomyosarcoma[6] variant that may lead to an incorrect therapy. Fetal rhabdomyoma is a well-circumscribed lesion, localized superficially in contrast with the rhabdomyosarcoma that presents infiltrated margins. They rarely show mitotic figures, present uniform cells, lack of nuclear hyperchromatism and no signs of necrosis. Fibrosarcomas and leiomyomas are part of the differential diagnosis. Treatment of fetal rhabdomyoma consists in local excision,[8] with a very low chances of recurrence if surgery was properly done.

We believe that this case is a very atipical location of the fetal rhabdomyoma in a 8 month old infant. Very few are described in the literature and it is very important to make an adequate differential diagnosis to prevent mistreatment.

REFERENCES

1. Helmberger RC, Stringer SP, Mancuso AA. Rhabdomyoma of the pharyngeal musculature extending into the prestyloid parapharyngeal space. AJNR 17: 1115-1118, 1996.
2. Enzinger FM, Weiss SW. Soft Tissue Tumors. 3rd ed. St. Louis, Mo, CV Mosby, pp. 523-536, 1995.
3. Dehner LP, Enzinger FM, Font RL. Fetal rhabdomyoma an analysis of nine cases. Cancer 30: 160-166, 1972.
4. Konrad EA, Meister P, Hübner G. Extracardiac rhabdomyoma report of different types with light microscopic and ultrastructural studies. Cancer 49: 898-907, 1982.
5. Kapadia SB, Meis JM, Frisman DM, Ellis GL, Heffner DK. Fetal rhabdomyoma of the head and neck. Hum Pathol 24: 754-765, 1993.
6. Di Sant´agnese PA, Knowles DM. Extracardiac rhabdomyoma: A clinicopathologic study and review of the literature. Cancer 46: 780-789, 1980.
7. Pérez-Alonso P, Sánchez-Simón R, Contreras F, Patrón –Romero M. Pathological case of the month. Arch Pediatr Adolesc 154: 1265-1266, 2000.
8. Gardner DG, Corio RL. Fetal rhabdomyoma of the tongue, with a discussion of the two histologic variants of this tumor. Oral Surg 56: 293-300, 1983.