Case Report

Multinodular adult rhabdomyoma in female: A rare case report

Shakti Kumar Yadav, Neelam Sood

Department of Pathology, Deen Dayal Upadhyay Hospital, New Delhi, India,

Abstract

Rhabdomyomas are rare benign mesenchymal tumors of striated muscle origin. These are classified as cardiac and extracardiac types. Extracardiac type is further classified as adult, fetal and genital types. Adult rhabdomyoma represents <2% of all muscular tumors. It mostly occurs in adults (median: 60 years). Males are more commonly affected (M:F = 3:1) and usually present as solitary lesion. We report a rare case of multinodular adult rhabdomyoma arising from the floor of the mouth. A 55-year-old female presented with a painless, soft, mobile, nontender and multinodular swelling in the right submandibular region and the floor of the mouth for 2 months. Fine-needle aspiration cytology showed cellular smears of large elongated to round cells with abundant eosinophilic granular cytoplasm, small nuclei and occasionally prominent nucleoli. Histology revealed partially encapsulated lesion showing sheets of large, oval to polygonal cells with abundant deeply eosinophilic and granular cytoplasm, with small peripherally placed nuclei with few cells showing prominent nucleoli. Prominent cytoplasmic vacuolations (periodic acid–Schiff positive) were present. The cells were positive for desmin and myogenin. This case is an extremely rare presentation of multinodular adult rhabdomyoma in a female. The correct identification of this lesion is important to avoid an unnecessarily aggressive resection, yet providing potentially curative therapy.

Keywords: Intraoral, rhabdomyoma, rhabdomyosarcoma

Address for correspondence: Dr. Neelam Sood, Head of the Department, Deen Dayal Upadhyay Hospital, New Delhi, India.
E-mail: neesu1234@hotmail.com
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INTRODUCTION

Rhabdomyomas are rare benign mesenchymal tumors of striated muscle origin. These are topographically classified as extracardiac and intracardiac types. Extracardiac type is very rare lesions with propensity toward the head-and-neck region. In contrast to intracardiac type, they are never associated with other malformations while the latter is often associated with tuberous sclerosis or other genetic diseases.[1]

Extracardiac type is further classified into adult, fetal and genital types. The adult and fetal types are typically seen in the head-and-neck region of the elderly and pediatric age groups, respectively. The genital type mostly presents as vulvovaginal lesion in middle-aged females.[2]

The adult type also known as rhabdomyomatous hamartoma or rhabdomyoma purum is the most common type of extracardiac rhabdomyoma, but represents <2% of all muscular tumors. It mostly occurs in adults older than 40 years (median: 60 years). Males are more commonly affected than females (3:1).[2-4] Majority of these tumors are solitary; however, cases of multinodular adult rhabdomyoma are also reported in previous literature.[2,5,6]

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Till date, only three cases of females with this lesion are reported in the English literature, of which only one case had multifocal. This is a rare case of multinodular adult rhabdomyoma originating from the floor of the mouth.

CASE REPORT

A 55-year-old female patient presented to the otorhinolaryngology department with complaints of painless swelling in the right submandibular region and the floor of the mouth for the last 2 months. It was soft, mobile, nontender and multinodular. The provisional diagnosis of salivary gland lesion was made, and the patient was referred to fine-needle aspiration cytology (FNAC) clinic for further evaluation.

Cytomorphology of the lesion revealed cellular smears showing large elongated to round cells with abundant eosinophilic granular cytoplasm, small nuclei and occasionally prominent nucleoli. Few cells showed cytoplasmic vacuolations and were positive for periodic acid–Schiff (PAS) staining. Immunostaining for myogenin was also present in the cells. Cytological diagnosis of rhabdomyoma was rendered followed by excision of the lesion and histopathological examination [Figure 1].

Grossly, four well-circumscribed, unencapsulated, nodular soft-tissue pieces were received ranging in size from 5 cm to 1 cm in maximum diameter [Figure 2a]. Microscopically, the lesion composed of large, oval to polygonal cells with the abundant amount of deeply eosinophilic and granular cytoplasm, with small peripherally placed nuclei with few cells showing prominent nucleoli; occasional giant cells were seen [Figure 2b-d]. Partial fibrous capsule and normal muscle was also seen. Prominent cytoplasmic vacuolations were present in fair number of cells which were found to be PAS positive [Figure 3a]. Immunohistochemically, the cells showed strong reactivity for myogenin (clone F5D, Dako Cytomation) and desmin (clone D33, Biocare Medical) [Figure 3b and c].

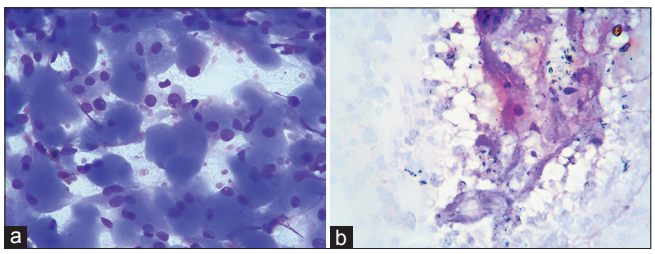

Figure 1: (a) Large round to oval cells with abundant cytoplasm and peripherally place small nuclei with prominent nucleoli (MGG, ×400), (b) immunoreactivity for myogenin (DAB, ×400)

Based on the above findings, the final histological diagnosis of adult rhabdomyoma was rendered.

DISCUSSION

Adult rhabdomyomas are rare benign skeletal muscle tumors. They are considered to be true neoplasms unlike cardiac rhabdomyomas, which are regarded as hamartomas.[6] These are most common in the head-and-neck region. This predilection is because tumor originates from the branchial musculature of the third and fourth branchial arches. Most of the reported cases are males in the age group of 40 years.[4]

The patient usually presents with a slow-growing asymptomatic painless, mobile, smooth, nodular mass and, sometimes, presents with globus sensation, hoarseness and dysphagia. Commonly reported locations are floor of mouth, soft palate, tongue and buccal mucosa.[3,7]

Radiological shows nonspecific well-demarcated mass having signal-intensity isointense or slightly hyperintense to that of skeletal muscle on T1- and T2-weighted magnetic resonance imaging. On computed tomography scan, it can show indistinct borders and mimic malignant tumor and lymphomas.[6,8]

Depending on the location of tumor, the differential diagnosis may include neurogenic or vascular tumors, oncocytoma, granular cell tumor and rhabdomyosarcoma. Sometimes, these lesions may be mistaken for malignant tumor of minor salivary glands when present in parapharyngeal space.[6,9]

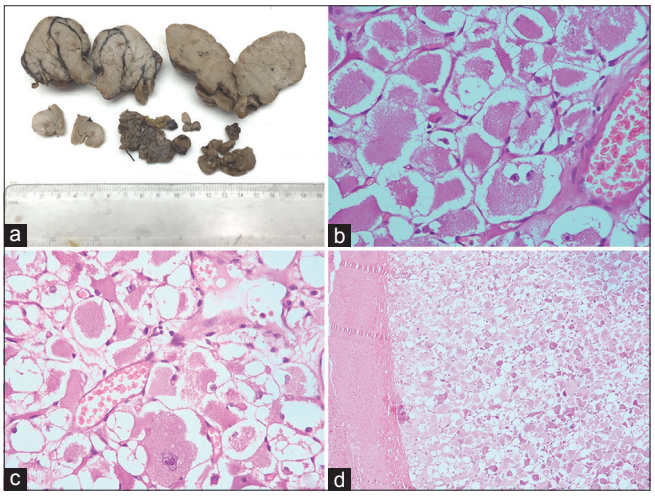

Figure 2: (a) Four nodular swellings showing gray tan cut surface and small salivary gland tissue piece, (b) tumor tissue composed of large round to polygonal cell having abundant eosinophilic granular cytoplasm (H and E, ×400), (c) occasional giant cells (H and E, ×400) and (d) partial capsule is identified at places (H and E, ×100)

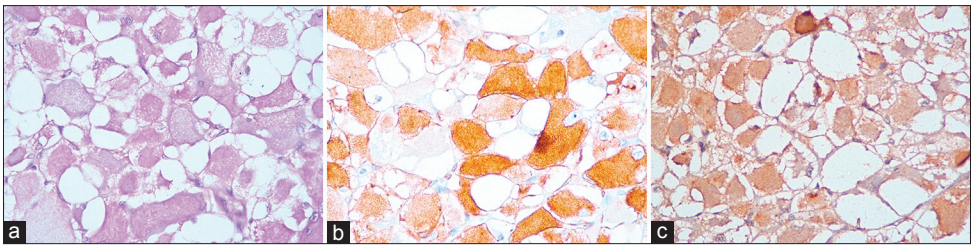

Figure 3: (a) Cell shows positivity for periodic acid–Schiff due to the presence of glycogen (periodic acid–Schiff, ×400), (b) strong immunostaining for desmin (DAB, ×400) and (c) myogenin (DAB, ×400) shows strong positivity

Macroscopically, the lesions are unencapsulated, well-circumscribed, deep tan to brown-colored, nodular or lobular mass.[3] Microscopically, adult rhabdomyoma is characterized by sheets of large polygonal to round cells with abundant deeply eosinophilic, granular cytoplasm. Cells have one or two peripherally placed vesicular nuclei; occasionally, prominent nucleoli can be present. Cytoplasmic vacuolization is present as in this case in many cells due to intracytoplasmic glycogen accumulation. Cytoplasm of some cells (spider cells) shows small acidophilic mass connected by thin strands to rim of peripherally condensed cytoplasm. Cross-striations and rod-like or jackstraw-like intracytoplasmic crystalline structure are usually easily identified although more evident on phosphotungstic acid hematoxylin staining. Mitoses and necrosis are absent. The cells show strong PAS positivity.[2,7]

Immunohistochemically, the cells show strong positivity for skeletal muscle markers such as muscle-specific myoglobin, actin, desmin and myogenin. Desmin is the most reliable marker for skeletal and smooth muscle differentiation, and it is also present in primitive and mature cells. Vimentin is present in primitive, but not in the mature skeletal muscle. Myoglobin is present in much greater quantity than in the fetal muscles, however, there is variable immunoreactivity for vimentin, S100 and smooth muscle actin. Tumor cells do not express cell proliferation markers such as Ki67 and proliferating cell nuclear antigen, which indicates that lesion is more likely hamartomas.[1,6] Electron microscopic examination shows longitudinal and transversely arranged myofibrils along with mitochondria and asterisk-shaped cytoplasmic inclusions.[2]

The microscopic diagnostic features of adult rhabdomyoma are fairly distinct in spite of its rarity. The differential diagnoses for this tumor include granular cell tumor, rhabdomyosarcoma, hibernoma, oncocytoma, paraganglioma and crystal-storing histiocytosis.[10]

Tumors in which vacuolated cells are predominant with few cells with striations, then differential diagnoses of granular cell tumor and hibernoma should be considered. In granular cell tumor, the cells are usually pale having indistinct borders with the absence of vacuolation and cross-striations and show diffuse S100 immunoreactivity and negative for desmin. Hibernoma mostly found in the interscapular region shows frequent intracytoplasmic vacuoles and the presence of intracellular lipid. Tumor is composed of deeply eosinophilic granular cells containing cytoplasmic lipid droplets, but there is no skeletal muscle differentiation and no glycogen. Malignant tumor with rhabdoid differentiation such as rhabdomyosarcoma and malignant rhabdoid tumor shows characteristic nuclear pleomorphism and atypia.[10]

Reticulohistiocytoma is composed of intermingled deeply acidophilic histiocytes, fibroblasts, xanthoma cells, multinucleated giant cells and chronic inflammatory elements. Typically, there is no skeletal muscle differentiation and glycogen. Crystal-storing histiocytosis associated with lymphoplasmacytic neoplasms shows monoclonal lymphoplasmacytic infiltrate. Crystal-storing cells are histiocytes, positive for CD68 but are negative for skeletal muscle markers and S100 protein. Oncocytoma is composed of polyhedral cells with finely granular, eosinophilic cytoplasm. The cells stain for epithelial markers, but do not express skeletal muscle markers.[10]

In paraganglioma, the tumor cells are arranged in an organoid pattern and express neuroendocrine markers like neuron-specific enolase, synaptophysin and chromogranin. However, the cells in paraganglioma are negative for skeletal muscle markers like muscle specific actin.[4]

CONCLUSION

The present case is an extremely rare presentation of multinodular adult rhabdomyoma in a female. This case showed that definitive diagnosis is possible on FNAC with immunocytochemistry. It is important that an early correct identification is made to avoid an unnecessarily aggressive resection, yet providing potentially curative therapy.

Even though not much literature exists, we only recommend surgery of lesions that are symptomatic

or cosmetically disturbing. The occurrence of multiple lesions (multinodular and multilobulated) has to be considered because most likely postoperative persistence of tumor lobules is the reason for the high recurrence rates documented in the literature.

Consent

Written informed consent has been obtained from the patient for the publication of this article and associated images.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Maglio R, Francesco S, Paolo M, Stefano V, Francesco D, Giovanni R. Voluminous extracardiac adult rhabdomyoma of the neck: A case presentation. Case Rep Surg 2012;2012:984789.
2. Amelia Souza A, de Araújo VC, Passador Santos F, Ferreira Martinez E, de Menezes Filho JF, Soares de Araujo N, *et al*. Intraoral adult rhabdomyoma: A case report. Case Rep Dent 2013;2013:741548.
3. Fletcher CD; World Health Organization. International agency for research on cancer. WHO Classification of Tumours of Soft Tissue and Bone. 4th ed. Lyon: IARC Press; 2013. p. 468.
4. Weiss SW, Goldblum JR, Enzinger FM. Enzinger and Weiss's Soft Tissue Tumors. 5th ed. Philadelphia, PA: Mosby Elsevier; 2008. p. 1258.
5. Wenig B. Neoplasms of the oral cavity, nasopharynx, oropharynx and neck. Atlas of Head and Neck Pathology. 2nd ed. London, UK: Elsevier; 2007. p. 254-389.
6. de Trey LA, Schmid S, Huber GF. Multifocal adult rhabdomyoma of the head and neck manifestation in 7 locations and review of the literature. Case Rep Otolaryngol 2013;2013:758416.
7. Favia G, Lo Muzio L, Serpico R, Maiorano E. Rhabdomyoma of the head and neck: Clinicopathologic features of two cases. Head Neck 2003;25:700-4.
8. Leboulanger N, Picard A, Roger G, Garabedian EN. Fetal rhabdomyoma of the infratemporal fossa in children. Eur Ann Otorhinolaryngol Head Neck Dis 2010;127:30-2.
9. Helmberger RC, Stringer SP, Mancuso AA. Rhabdomyoma of the pharyngeal musculature extending into the prestyloid parapharyngeal space. AJNR Am J Neuroradiol 1996;17:1115-8.
10. Veziroglu F, Uçkan S, Sengüven B. Adult type rhabdomyoma in a child. Oral Oncol Extra 2006;42:213-6.