SPINDLE CELL RHABDOMYOSARCOMA OF THE TONGUE – A RARE ENTITY

MT Khadim
A Asif
Z Ali
S Jamal
R Ahmad

Department of Histopathology,
Armed Forces Institute of Pathology,
Rawalpindi, Pakistan

Correspondence to:
Dr. Zafar Ali
House 20, Street 11, Sector F 6/3,
Islamabad, Pakistan
E-mail: zafarali@hotmail.com

Spindle cell variant of Rhabdomyosarcoma is a rare malignant tumor in adults but even more so in tongue. We report a case of a 17 years old boy who presented with a polypoidal swelling on the tip of the tongue. The mass was excised and the specimen was sent for histopathological evaluation which showed a malignant spindle cell tumor. On the basis of morphological features and immunohistochemistry findings a final diagnosis of rhabdomyosarcoma, Spindle cell variant was made.

Key words: desmin, rhabdomyosarcoma, tongue

INTRODUCTION:

Rhabdomyosarcoma (RMS) comprises a group of soft tissue neoplasms that shares the propensity to undergo myogenesis.[1] There is a bimodal distribution of presentation with an initial peak incidence between 2-5 years of age and a second surge at 10-19 years.[2] This results in its resemblance with different stages of skeletal muscle development during prenatal life. The annual incidence of RMS is about 8 in a million children.[2] RMS is the most common sarcoma in infants and children and represents 5-15% of all the solid neoplasms.[2] Most common head and neck site include the orbit, nasopharynx, paranasal sinuses, cheek, neck, middle ear and larynx. The occurrence of RMS in the tongue is uncommon.[2] The spindle cell variant of rhabdomyosarcoma (RMS) is commonly encountered tumor in the paratesticular region of children.[3] Other sites if involved includes the oral cavity, parotid gland, nasopharynx and nasal cavity. Less than 10 % of adult rhabdomyosarcomas are spindle cell variant.[4-6] Histologically they show proliferation of spindle shaped cells with pale cytoplasm arranged in interlacing fascicles.[7] Individual cells have small nuclei and inconspicuous nucleoli. Scattered rhabdomyoblasts are seen. Only few case of spindle cell RMS of tongue have been reported in literature.[2,8]

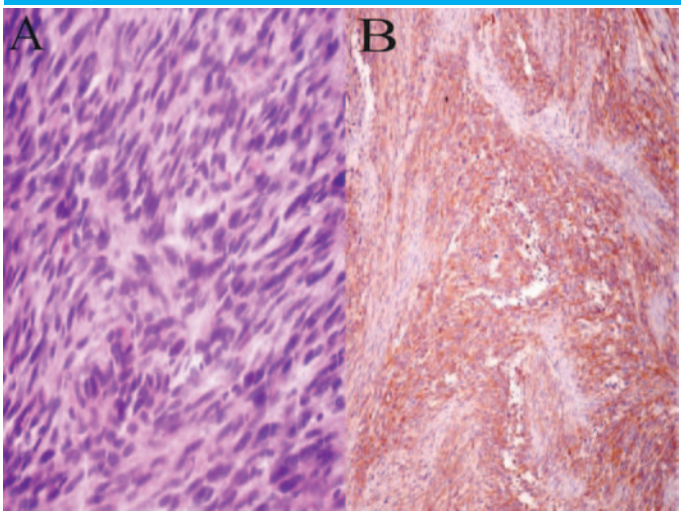
CASE REPORT:

We report a case of a 17 years old male who presented with a swelling on the tip of his tongue. The swelling had developed over a period of 8 months. The clinical impression was of Myoepithelioma. The lesion was surgically excised and the specimen was sent to the Histopathology Department for histological evaluation. On gross examination the specimen consisted of a yellow white nodular piece of tissue measuring 7 × 5 × 3 cm. On serial slicing the tumor had whorled appearance. Multiple sections of the tumor were taken and then examined. Microscopic picture revealed a tumor composed of fascicles of atypical pleomorphic spindle shaped cells . The cells were arranged in fascicles and focal storiform arrangement. Many typical rhabdomyoblasts having hyperchromatic eccentric nuclei and abundant eosinophilic cytoplasm were seen. The tumor cells were separated by fibrous stroma. Areas of tumor necrosis were also seen. The tumor had infiltrating borders and was involving the circumferential resection margin and invading the muscles. Immunohistochemical markers were applied. Desmin, Myogenin and Myo D1 were positive. Vimentin showed focal positivity. S-100, GFAP and CK AE1/AE3 were found to be negative. The morphological features and immunohistochemistry results favoured the diagnosis of rhabdomyosarcoma, spindle cell variant.

DISCUSSION:

Spindle cell RMS is a variant of embryonal RMS composed of elongated spindle cells and associated with a good prognosis.[7] Spindle cell variant is relatively newer entry, first described in 1992.[7] It is uncommon and usually found in the paratesticular region in children.[3] It is an unusual finding in adults and the tongue is a rare site.[4] Clinically these tumors may present as bulging, infiltrative, growing soft tissue masses which may be fungating exhibiting pressure effects with difficulty in speech and swallowing.[2] On gross examination of the resected specimens, the tumor has firm tan yellow color and whorled cut surface resembling leiomyoma. Histologically, the tumor cells present with shapes of different developmental stages of myogenesis.[1] Key cell to recognize by routine microscopy is the rhabdomyoblast, a cell with an eccentric round nucleus and variable amounts of brightly eosinophilic cytoplasm. Less than 30% of cases may show skeletal striations.[2] The mitotic count is usually low. RMS spindle cell variant being composed of elongate spindle cells arrayed in tight fascicles with variable amounts of intervening collagen. A group of these lesions were consequently described by Cavazanna et al.[7], who confirmed their superior prognosis and found that the majority occurred in the paratesticular region. Spindle cell rhabdomyosarcomas may arise in adults, in whom they may have a worse prognosis. According to the series by Nascimento and Fletcher, [4] they most commonly arose in the head and neck rather than paratesticular soft tissue. In the setting of adult neoplasia, they can pose a diagnostic dilemma because of their resemblance to smooth muscle tumors and other spindle cell lesions. This challenge can be solved by Immunohistochemistry and/or electron microscopic studies. Thus, immunohistochemistry is used extensively to distinguish RMS from its mimics.[1] Myogenin and MyoD1, myogenic transcriptional regulatory proteins expressed early in skeletal muscle differentiation, are considered sensitive and specific markers for RMS and are more specific than desmin and muscle-specific actin and more sensitive than myoglobin. Because the extent of myogenin expression in RMS is much greater than in non-RMS, it is a very useful marker when interpreted in the context of other clinicopathologic data. In our case the clinicopathological picture as well as the Immunohistochemical results corresponds to the ultimate diagnosis of spindle cell variant of Rhabdomyosarcoma of the tongue.

Fig: 1. Inset A : H/E (Haematoxylon and Eosin) – Spindle shaped cell arranged in interlacing fascicles and focal storiform pattern. Inset B : Immunohistochemistry for Myo D1 showing nuclear positivity



CONCLUSION:

Spindle cell variant of Rhabdomyosarcoma is a rare tumor of the tongue. It poses a diagnostic challenge due to its histological similarity with other smooth muscle cell neoplasms.

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