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Case Report

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NASOPHARYNGEAL RHABDOMYOSARCOMA: A CASE REPORT

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ABSTRACT

Introduction: Rhabdomyosarcoma is a soft tissue sarcoma arising from cells of myogenic lineage. Most common sites being extremities, perirectal and perineal region. It is a malignant tumor of soft tissue in children under 15 years of age, accounting 5% to 10% of all childhood malignancies. Head and neck region is frequently involved in childhood. For pediatric rhabdomyosarcoma cases, most of them have orbital primaries in the head and neck region, nasopharynx and nasal cavity cases are rarely seen. **Case Report:** An 11 years old boy presented with history of nasal mass, nasal obstruction and epistaxis. These symptoms started 4 to 5 months back and gradually progressed. The histological diagnosis of rhabdomyosarcoma was confirmed by immunohistochemistry method. We report this case because of its unique location in nasopharynx. **Discussion:** Rhabdomyosarcoma originates from immature mesenchymal cells that are committed to skeletal muscle differentiation. It is the most frequent soft tissue tumor in children. Histologically, it is distinguished in 3 types: embryonic, alveolar and pleomorphic. Embryonic type is mainly affect children less than 5 years of age. Pediatric rhabdomyosarcoma cases in head and neck region mainly have orbital primaries whereas nasopharynx and nasal cavity rhabdomyosarcoma cases are rarely seen. Only histology with immunohistochemistry provides specific diagnosis, in the form of diffuse desmin and myogenin staining. **Conclusion:** Rhabdomyosarcoma is a most common type of soft tissue sarcoma in children. It is localized in the head and neck region in 40% cases. Treatment of rhabdomyosarcoma is complex, including multi-drug chemotherapy, radiotherapy and surgery.

KEYWORDS: Rhabdomyosarcoma, Nasopharynx, Myogenin.

INTRODUCTION

Rhabdomyosarcoma is a soft tissue sarcoma arising from the cells of myogenic lineage. Most common sites being extremities, perirectal and perineal region.[1] It is a malignant tumor of soft tissue in children under 15 years of age, accounting for 5% to 10% of all childhood malignancies. Head and neck region is frequently involved in childhood. For pediatric rhabdomyosarcoma cases, most of them have orbital primaries in the head and neck region, nasopharynx and nasal cavity cases rarely seen. Histologically, it is distinguished into 3 types: embryonic, alveolar and pleomorphic.

CASE REPORT

A healthy 11 years old male child presented with a complaint of nasal obstruction and epistaxis in outpatient department of Otorhinolaryngology at Pt. B. D. Sharma PGIMS, University of Health Sciences, Rohtak. These symptoms started 4 to 5 months back and are gradually progressing. On physical examination a nasal mass was observed. Magnetic resonance imaging of head and neck region showed a mass in nasopharynx which was extending into nasal cavity. Biopsy samples were obtained from the nasal mass and send to the department of Pathology for histopathological examination, the excised specimen consisted of five grayish white to grayish brown soft tissue pieces measuring together 0.8X0.6X0.3 cm. Microscopic examination of hematoxylin-eosin stained sections revealed marked cellularity, sheets of small round cells with eosinophilic cytoplasm, high nucleo-cytoplasmic ratio. Cells had hyperchromatic, pleomorphic nuclei and conspicuous nucleoli [Figure 1]. On immunohistochemistry the tumor cells were vimentin positive, desmin positive, myogenin positive and cytokeratin negative consistent with diagnosis of embryonic type of rhabdomyosarcoma [Figure 2A, 2B, 2C].

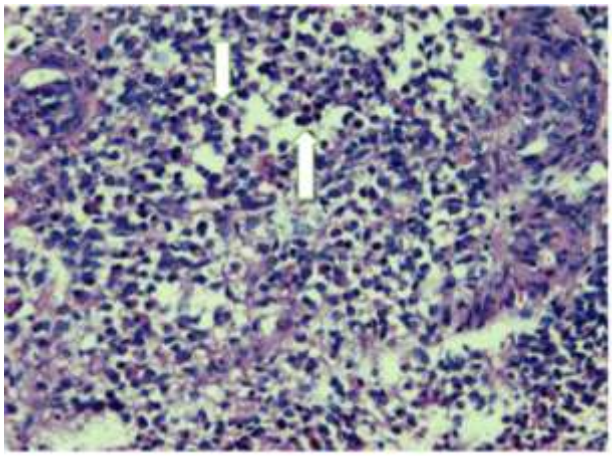
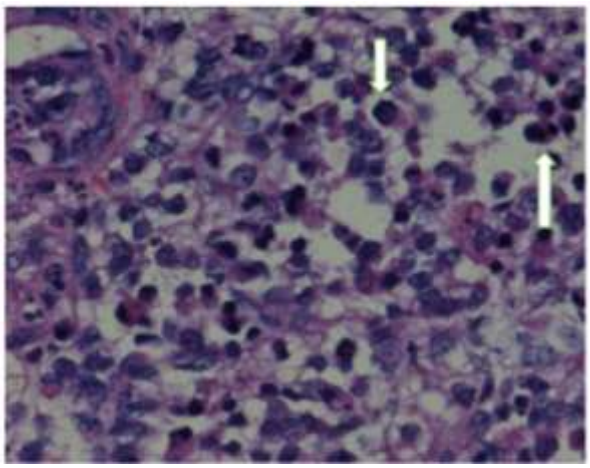


Figure 1: An embryonal rhabdomyosarcoma showing marked cellularity, sheets of small round cells with eosinophilic cytoplasm, high nucleo-cytoplasmic ratio [white arrows]. Cells having hyperchromatic, pleomorphic nucleus and conspicuous nucleoli. (hematoxylin-eosin; magnification 200X).



(Hematoxylin-eosin stained slide showing marked cellularity, sheets of small round cells with eosinophilic cytoplasm, high nucleo-cytoplasmic ratio [white arrows]. Cells having hyperchromatic, pleomorphic nucleus and conspicuous nucleoli. Embryonal rhabdomyosarcoma; magnification 400X)

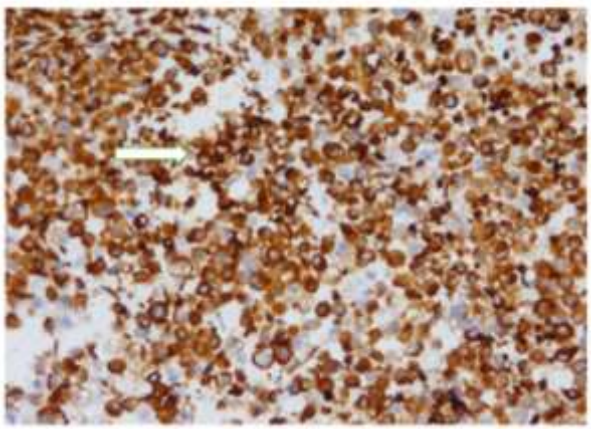


Figure 2A: Tumor cells showing vimentin positivity [white arrow]; magnification 200X).

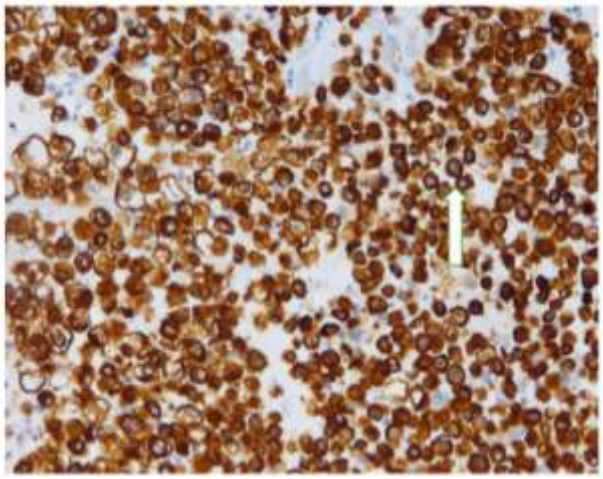


Figure 2B: Tumor cells showing desmin positivity [white arrows] magnification 200X.

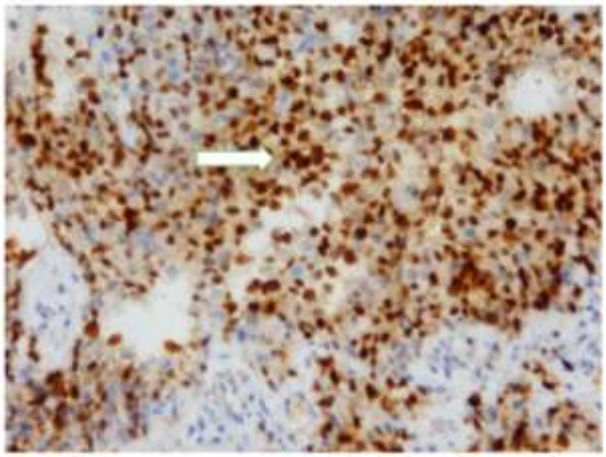


Figure 2C: Tumor cells showing myogenin positivity; [white arrow] magnification X 200.

DISCUSSION

Rhabdomyosarcoma originates from the immature mesenchymal cells that are committed to skeletal muscle differentiation. Embryonic type mainly affecting children less than 5 years of age. Embryonal type is the most common histological type of the head and neck rhabdomyosarcoma.[2] For pediatric rhabdomyosarcoma cases, most of them have orbital primaries in the head and neck region, nasopharynx and nasal cavity cases rarely seen. Treatment of pediatric rhabdomyosarcoma often requires a multimodal therapy which include surgery, chemotherapy and radiotherapy. Surgery is the cornerstone of this combined treatment. Except orbital primaries, head and neck rhabdomyosarcoma cases must be treated with surgery, with total excision if possible. The most commonly used regimens include vincristine and actinomycin-D in combination with alkylating agents such as cyclophosphamide or ifosfamide. Embryonal (botryoid) variant has a good prognosis. 5 years survival is 81%. Only histopathology with immunohistochemistry provides specific diagnosis, in the form of diffuse desmin and myogenin staining.

CONCLUSION

Pediatric rhabdomyosarcoma cases, most of them have orbital primaries in the head and neck region, nasopharynx and nasal cavity cases rarely seen. Embryonic type of rhabdomyosarcoma is most common and mainly affect the children less than 5 years of age. Embryonic type is associated with best prognosis out of the other types. Treatment of rhabdomyosarcoma is complex which include chemotherapy, radiotherapy and surgery.

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