www.jmscr.igmpublication.org

Impact Factor 3.79
ISSN (e)-2347-176x
crossref DOI: http://dx.doi.org/10.18535/jmscr/v3i9.05

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Alveolar Rhabdomyosarcoma Arising From Maxillary Antrum in a Adult: A Rare Case Report

Authors
Amod Kumar[1], Anju Singh[2] , Sanjeet Kumar Singh[3], M.A. Muzaffar[4], Subhash Chandra Jha[5], Shahab Raza[6] Rakesh Kumar Singh[7]
[1]Consultant Pathologist, SRL Diagnostics, Patna , [2,3]Assistant Professor, Department of Pathology, Indira Gandhi Institute of Medical sciences, Patna[4]Tutor, Department of Pathology, Patna Medical College, Patna[5] Assistant Professor, Department of Pathology, Government Medical College, Bettiah[4]Assistant Professor, Department of Pathology, Indira Gandhi Institute of [6]Associate Professor, Dept of Pathology, Patna Medical College, Patna,[7]Additional Professor, Dept of ENT, Indira Gandhi Institute of Medical Sciences, Patna
Corresponding Author
Dr. Subhash Chandra Jha
Assistant professor, Department of Pathology, Government, Medical College, Bettiah, Pin code: 845438,
Email ID: *jhapatho25@gmail.com Mobile: 09771434052*

Abstract
Rhabdomyosarcoma is one of the most common soft-tissue sarcomas in children under 15 years of age, and the head and neck is the principal location for childhood rhabdomyosarcoma; however head and neck rhabdomyosarcoma is rare in adults, occasionally occurs in nasal cavity and paranasal sinuses, it is rare at this site in adults. The typical subtypes of embryonal, alveolar and pleomorphic rhabdomyosarcoma are diagnosed histopathologically. The markers connoting simple myogenous or skeletal muscle differentiation has contributed tremendously to the diagnosis of even poorly differentiated rhabdomyosarcoma .The alveolar variant is more commonly seen in extremities and is very unusual in nasal and paranasal sinuses. Desmin and myogenin were highly positive while others were negative. Thus, a rare diagnosis of alveolar rhabdomyosarcoma arising from maxillary antrum was confirmed. We present a rare case of Alveolar rhabdomyosarcoma at unusual site in an adult.

Keywords: *Rhabdomyosarcoma, alveolar, adult, paranasal sinus*

Introduction

Rhabdomyosarcoma (RMS) is histologically and genetically heterogeneous malignant tumor of skeletal muscle origin.[1] It is a heterogeneous group of tumors with respect to their molecular basis, degree of differentiation, histopathology and clinical behavior.[2] The typical subtypes of embryonal, alveolar and pleomorphic rhabdomyo-sarcoma are diagnosed histopathologically. The availability of markers connoting simple myogenous or skeletal muscle differentiation has contributed tremendously to the diagnosis of even poorly differentiated rhabdomyosarcoma .[3] The alveolar variant is more commonly seen in extremities and is very unusual in nasal and paranasal sinuses.[4] The common histopathological differential diagnoses are alveolar soft part sarcoma, malignant round cell and clear cell

tumors. The immunohistochemical positivity of desmin is very useful and myogenin positivity confirms diagnosis of rhabdomyosarcoma in poorly differentiated type.Myogenin expression determines commitment and differentiation of primitive mesenchymal cells into skeletal muscle. The expression of myogenin has been demonstrated to be extremely specific for rhabdomyoblastic differentiation; therefore very useful marker in the differential diagnosis of rhabdomyosarcoma from other malignant small round cell tumors of childhood.[5] Desmin and myogenin are highly positive in alveolar variant. It is one of the most common soft-tissue sarcomas in children under 15 years of age and the head and neck is the principal location for childhood rhabdomyosarcoma, however head and neck rhabdomyosarcoma is rare in adults.[6, 7, 8]

Case Summary:

A 30 year adult female presented with bilateral nasal mass, epistaxis and proptosis of left eye (Fig.1). Her bilateral cervical lymph nodes were enlarged. High resolution computed tomography (HRCT) of paranasal sinuses and orbits showed a 5.3 x 4.7 cms large lobulated heterogeneously enhancing left maxillary antral mass extending into nasal cavity, nasopharynx, ethmoidal and frontal sinus with erosion of cribriform plate and pushing the left orbital structures but not infiltrating any of orbital(Fig.2) . Fine needle aspiration cytology (FNAC) from mass and lymph nodes showed predominantly dispersed round cells , few stripped nuclei and multinucleated giant cells with tendency to form rosette-like structure at places (Fig.3A,C). Individual cells had eccentric and hyperchromatic nuclei showing features suggestive of round cell tumor possibly malignant round cell tumor. Histopathology of biopsy from left nasal mass showed nests of round to oval tumor cells separated by connective tissue septa. Few cells tend to detach giving an alveolar appearance at places (Fig.3B). Tumor cells had hyperchromatic nuclei and acidophilic cytoplasm. Occasional multinucleated giant cells are also noted. Diagnosis of malignant round cell tumor likely Alveolar rhabdomyosarcoma was made. Markers for CD 45, CD99, cytokeratin (CK), desmin and myogenin were used for immunohistochemical staining. Desmin and myogenin were highly positive (Fig.4A, 4B). CD45, CD99 and cytokeratin were negative (Fig.5A, B, C) .Thus, a rare diagnosis of alveolar rhabdomyosarcoma arising from maxillary antrum was confirmed.

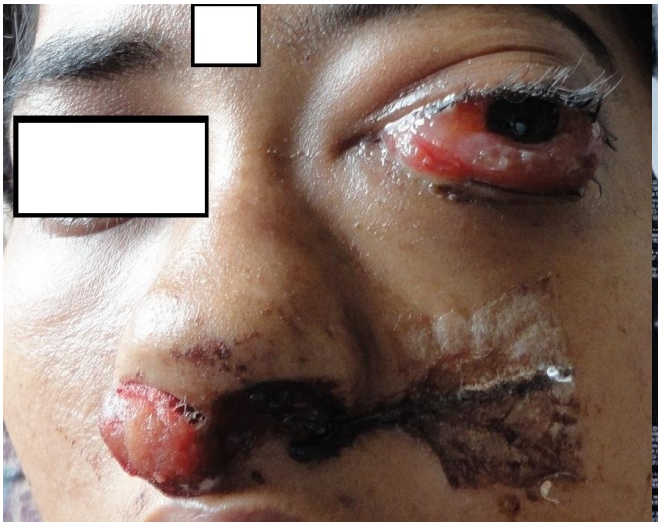

Fig.1. Adult female showing left maxillary antral mass extending into nasal cavity, nasopharynx, and left eye.

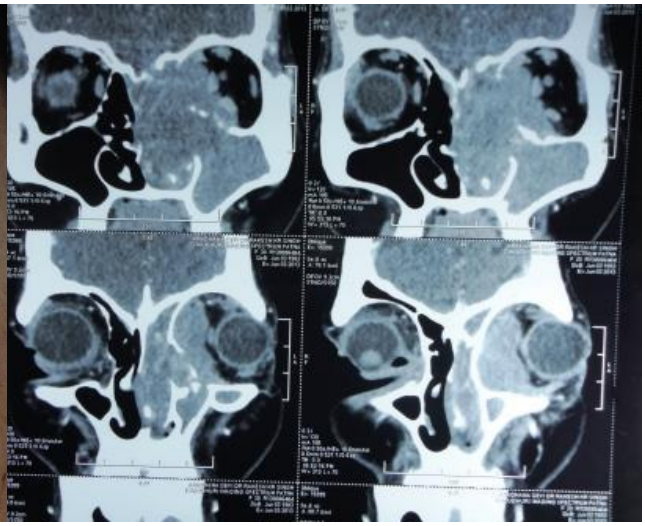

Fig. 2. Computed tomography showing left maxillary antral mass extending into nasal cavity, nasopharynx, ethmoidal and frontal sinus with erosion of cribriform plate

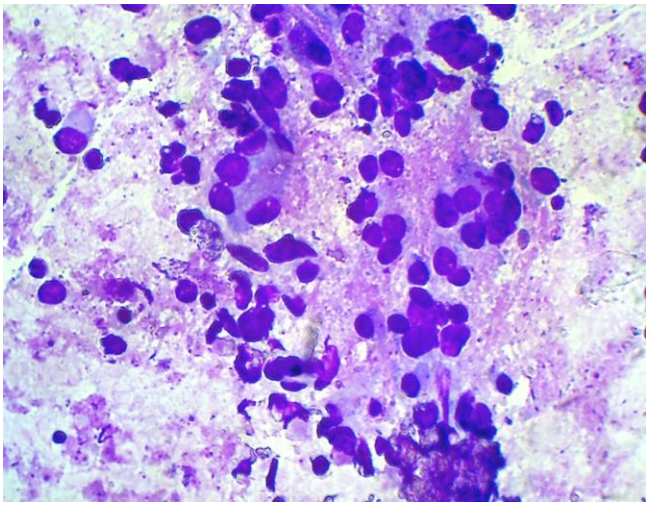

Fig. 3A. Smear from nasal mass showing round to oval tumor cells

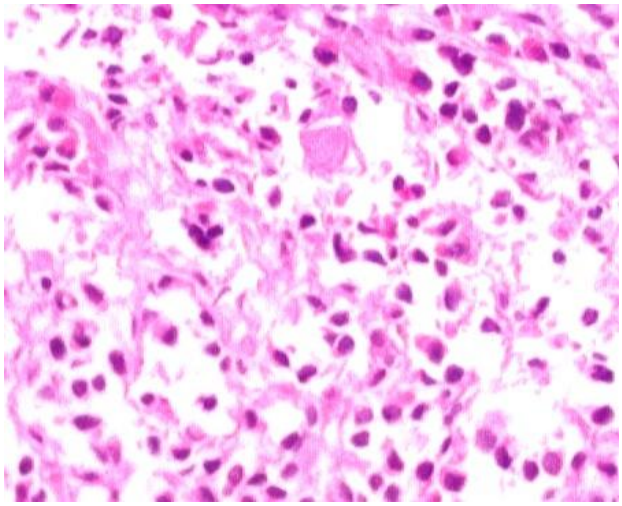

Fig. 3B. Histopathology showing round to oval tumor cells arranged in alveolar pattern

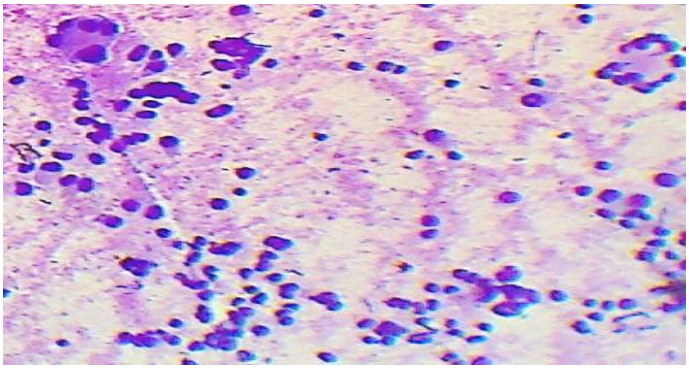

Fig. 3C.Smear from lymph node showing tumor cells in the background of lymphocytes

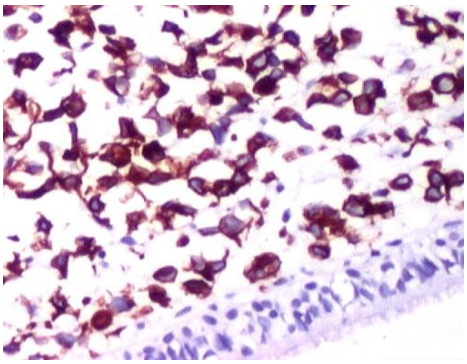

Fig. 4A. IHC showing cytoplasmic positivity for Desmin.

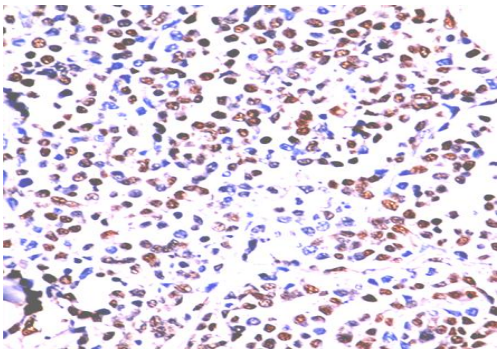

Fig. 4B. IHC showing nuclear positivity for Myogenin.

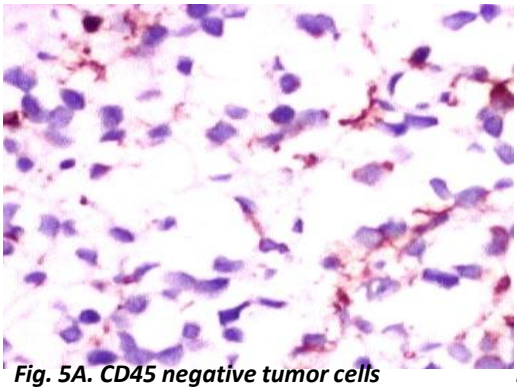

Fig. 5A. CD45 negative tumor cells

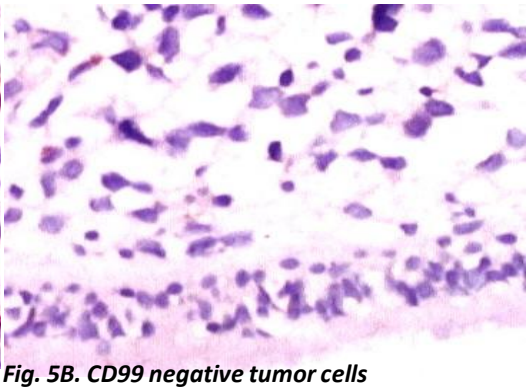

Fig. 5B. CD99 negative tumor cells

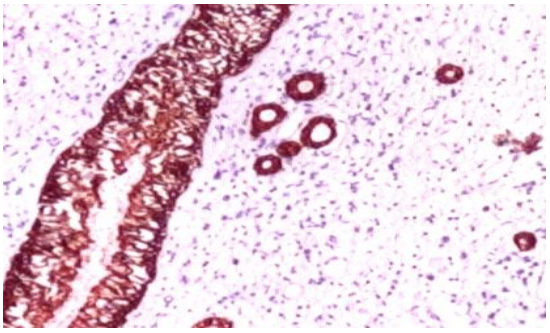

Fig.5C CK is negative in tumor cells but positive in ductal cells.

Discussion

Alveolar rhabdomyosarcoma is less frequent and accounts for 10% to 20% of all rhabdomyosarcomas.[3] It affects chiefly children and young adults between 10 and 25 years of age and occurs more frequently in the extremities; the most common locations being forearms, arms, perirectal and perineal regions.[1] Adult rhabdomyosarcomas do not show a male preponderance, as is noted in paediatric patients.[8] It occurs with a predilection in extremities, while paediatric rhabdomyosarcomas occur predominantly in head and neck sites. [7, 8] The head and neck is extremely rare location of adult rhabdomyosarcoma, and five- year survival rate is less than 8% in these cases.[8] Rhabdomyosarcoma of paranasal sinuses accounts for 10% to 15% of adult head and neck rhabdomyosarcomas.[6] These are often advanced and locally invasive, and the ethmoidal or maxillary sinus is the most common site affected.[4] Lung and regional lymph nodes are the most common metastatic sites of alveolar rhabdomyosarcoma.[1] .Along with tumor invasiveness, metastasis and regional lymph node involvement, age at diagnosis is a predictor of outcome.[4] Overall survival is worse for adults than for children.[4] Alveolar rhabdomyosarcomas are associated with translocation t(2;13)(q35;q14) or t(1;13)(p36;q14).[1,2.]

Conclusion

Rhabdomyosarcoma is a well-recognized tumor in children and occasionally occurs in nasal cavity and paranasal sinuses. The alveolar variant is more commonly seen in extremities and is very unusual in nasal and paranasal sinuses.

References

1. Rosai and Ackerman's Surgical Pathology. 10th ed. 2011, pp. 2166-2171.
2. Enzinger and Weiss's Soft tissue tumors. 4th ed. 2001, pp. 785-828.
3. Dias P, Chen B, Dilday B, Palmer H, Hosoi H, Singh S et al. Strong Immunostaining for Myogenin in Rhabdomyosarcoma Is Significantly Associated with Tumors of the Alveolar Subclass . Am J Pathol 2000; 156(2):399-408.
4. Jeong Hoon Lee, Mi Sook Lee, Byung Hee Lee, et al. Rhabdomyosarcoma of the head and neck in adults: MR and CT findings.

AJNR Am J Neuroradiol 1996;17:1923-28.

5. Kumar S, Perlman E, C A Harris, Raffled M ,Tsokos M .Myogenin is a Specific Marker for Rhabdomyosarcoma: An Immunohistochemical Study in Paraffin-Embedded Tissues**.** Mod Pathol 2000;13(9):988–993
6. Nayar RC, Prudhomme F, Parise O Jr, Gandia D, Luboinski B, Schwaab G. Rhabdomyosarcoma of the head and neck in adults: a study of 26 patients. Laryngoscope 1993;103:1362-66.
7. Feldman BA. Rhabdomyosarcoma of the head and neck. Laryngoscope 1982;92:424-40.
8. Tsung-Han Wu, Jen Seng Huang, Hung-Ming Wang, Cheng-Hsu Wang, Kun-Yun Yeh. Long term survivors of adult rhabdomyosarcoma of maxillary sinus following multimodal therapy: Case reports and literature reviews. Chang Gung Med J 2010;33:466-471.

s