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

Authors
Amod Kumar¹, Anju Singh², Sanjeet Kumar Singh³, M.A. Muzaffar⁴, Subhash Chandra Jha⁵, Shahab Raza⁶ Rakesh Kumar Singh⁷

¹Consultant Pathologist, SRL Diagnostics, Patna 2,3Assistant Professor, Department of Pathology, Indira Gandhi Institute of Medical sciences, Patna ⁴Tutor, Department of Pathology, Patna Medical College, Patna ⁵Assistant Professor, Department of Pathology, Government Medical College, Bettiah ⁶Assistant Professor, Department of Pathology, Indira Gandhi Institute of ⁷Associate Professor, Dept of Pathology, Patna Medical College, Patna 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 rhabdomyosarcoma 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

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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. 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.

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

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
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