

Nasal Alveolar Rhabdomyosarcoma

Dr. Bishal Datta¹, Dr. Manisa Mohanty², Dr. Prasana Kumar Satpathy³,
Dr. Raghmani Mohanty⁴

^{1,2,3,4}(Department of Pathology, Hi-Tech Medical College & Hospital/Utkal University, Bhubaneswar)

Abstract: Rhabdomyosarcoma is a soft tissue sarcoma arising from cells of myogenic lineage. They can be divided into Embryonal rhabdomyosarcoma (ERMS), Alveolar rhabdomyosarcoma (ARMS) and Pleomorphic rhabdomyosarcoma. ARMS occurs in 10-25 years age group and the most common sites being extremities, perirectal and perineal regions, head and neck region is frequently involved in childhood but is exceedingly rare in adults. A female aged about 55 years presented in Otolaryngology department with history of left sided nasal obstruction for about 6 months. The excised specimen was sent to our Department of Pathology for histopathological examination. ARMS is an aggressive condition and carries a distinctly worse prognosis. We report the case due to the uniqueness of the age and location of the tumor.

Keywords: Alveolar rhabdomyosarcoma (ARMS), nasal cavity.

I. Introduction

Rhabdomyosarcoma (RMS) are skeletal muscle tumors under the soft tissue sarcoma category arising from cells of myogenic lineage. They can be divided into Embryonal rhabdomyosarcoma (ERMS), Alveolar rhabdomyosarcoma (ARMS) and Pleomorphic rhabdomyosarcoma (PRMS).

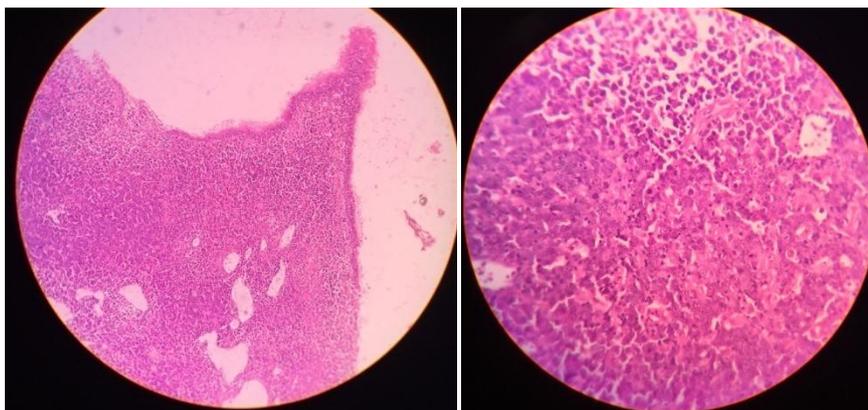
ARMS occur in 10-25 years age group, with a predilection for men. The most common sites being extremities, perirectal and perineal regions, head and neck region are frequently involved in childhood but are exceedingly rare in adults. ^[1] As a proportion of rhabdomyosarcomas as a whole, rhabdomyosarcomas in adults account for no more than 10%. ^[2]

II. Case History

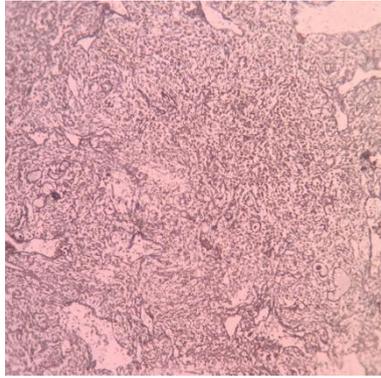
A female aged about 55 years presented in the Otolaryngology department with history of left sided nasal obstruction and epistaxis for about 6 months. On examination, single, spherical, pink mass about 2 cm x 1 cm in diameter was seen in left nasal cavity. The right nasal cavity and the oral cavity were normal. There were no palpable cervical lymph nodes.

The routine pre-operative profile was normal. The excised specimen was sent to our Department of Pathology for histopathological examination.

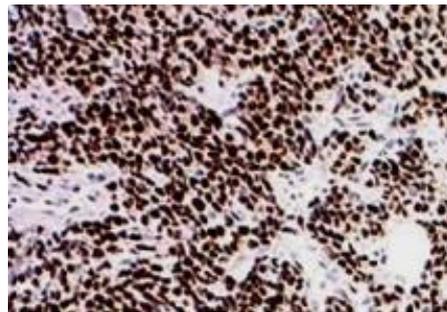
Grossly, there were multiple bits of grayish brown tissue all together measuring 2 cm X 1 cm. The section from the nasal mass showed tumor lined by respiratory epithelium and surrounded by wide areas of necrotic tissue. The tumor cells are round, oval to spindle shaped cells with a few cells showing abundant eosinophilic cytoplasm with eccentric nuclei. Cells are arranged in alveolar pattern and in sheets. Cells had hyperchromatic, pleomorphic nuclei and conspicuous nucleoli. Tumor is surrounded by lymphoplasmacytic infiltration.



We performed a reticulin stain which revealed the alveolar pattern of arrangement around each cell.



We also carried out an immunohistochemical study for myogenin, which stained positive on our prepared sections.



III. Conclusion

The first description of rhabdomyosarcoma was by Weber in 1854. ^[3] However, the “definitive” publication is usually considered to be by Stout in 1946, 92 years later. ^[4]

About 7 weeks into the development of an embryo, cells called rhabdomyoblasts (which will eventually form skeletal muscles) begin to form. These are the cells that can develop into rhabdomyosarcoma. Because this is a cancer of embryonal cells, it is much more common in children, although it does sometimes occur in adults. ^[5]

Alveolar rhabdomyosarcoma is the second most common subtype, accounting for approximately 31% of all rhabdomyosarcomas. ^[6]

ARMS most often occurs in large muscles of the trunk, arms, and legs. The cells of ARMS look like the normal muscle cells seen in a 10-week-old fetus. ^[7]

Most RMS develops in children, but adults can also have the condition. Adults are more likely to have faster-growing types of RMS and they have them in parts of the body that are difficult to treat. Because of this, RMS in adults is often harder to treat effectively. Some rare inherited conditions have been associated to increase the risk of RMS like Li-Fraumeni syndrome, Beckwith-Wiedemann syndrome, Neurofibromatosis type I, Costello syndrome and Noonan syndrome.

Immunohistochemistry has showed to be of great worth. Barely, there is any other condition which has such wide collection of markers. Of the panel, the utmost importance is myogenin. The myogenin gene codes for a phosphoprotein that induces skeletal muscle differentiation in mesenchymal cells. The protein, which has a high degree of specificity, can be demonstrated in the nuclei of the tumor cells in all types and virtually all cases of rhabdomyosarcoma, but it is expressed in a particularly strong and widespread fashion in the alveolar type. Probably because of this very fact, diffuse expression of myogenin is a marker of poor prognosis. ^[1]

ARMS carries a reproducible tumor specific chromosomal translocation, t(2;13)(q35;q14), resulting in PAX3-FOXO1 fusion. ^[2]

ARMS is an aggressive condition. It is more prone to metastasis to the lung and regional lymph node and carries a distinctly worse prognosis. Surgical excision is the treatment of choice. Also, multiagent chemotherapy with judicious surgery can be line of treatment for selected patients.

References

- [1]. Juan Rosai. Soft tissues. Rosai and Ackerman's surgical pathology (11th edition). Elsevier Saunders. 2012
- [2]. Christopher D.M. Fletcher. Diagnostic histopathology of tumors (4th edition, vol 2). Elsevier Saunders.2013
- [3]. Weber, CO. Anatomische Untersuchung Einer Hypertrophischen Zunge nebst Bemerkungen uber die Nubildung querquestreifter Muskelfern, Virchow Arch. Pathol Anat. 7:115, 1854.
- [4]. Stout AP: Rhabdomyosarcoma of the skeletal muscles, Ann Surg 1946; 123: 447-472.
- [5]. Cancer.org. 2014. Available from <http://www.cancer.org/acs/groups/cid/documents/webcontent/003136-pdf.pdf>
- [6]. Sharon WW, John RG. Enzinger and Weiss's soft tissue tumors (5th edition). Mosby Elsevier 2008
- [7]. Cancer.org. 2014. Available from <http://www.cancer.org/acs/groups/cid/documents/webcontent/003136-pdf.pdf>.