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Case Report

Metastatic deposit of Alveolar rhabdomyosarcoma in cervical lymphadenopathy in a 23 year male patient: A case report

Shana Nikhat Khan[1], Ranjana Hawaldar[1]*, Ekta Patidar[1]

[1]Dept. of Pathology, Sampurna Sodani Diagnostic Clinic, Indore, Madhya Pradesh India

Abstract

Cervical lymphadenopathy due to metastatic alveolar rhabdomyosarcoma (ARMS), presenting without a recognizable primary tumour, is a rare entity. We report a case of 23 year male patient diagnosed with fine needle aspiration, histopathology and IHC (Immunohistochemistry) markers for definitive diagnosis. The immunohistochemical analysis revealed intense positivity for myogenin and desmin, favouring the diagnosis of alveolar rhabdomyosarcoma.

Keywords: Rhabdomyosarcoma, Immunohistochemistry, Alveolar rhabdomyosarcoma.

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1. Introduction

Alveolar rhabdomyosarcoma (ARMS) is a primitive malignant high grade neoplasm characterized by round cells and partial differentiation of skeletal muscles.[1] Alveolar rhabdomyosarcoma (ARMS) occur at all ages, more often in adolescents and young adults than in younger children. The median age of occurrence is between 6.8 and 9.0 years.[2] It is difficult to diagnose as it represents only 3% of all soft tissue sarcoma in adults. Rhabdomyosarcoma (RMS) of head and neck of adults has a poor prognosis. Rhabdomyosarcoma is a common soft tissue tumour in children but rare in adults and its therapeutic management remains non-standardized.

2. Case Report

The patient was a 23 year old male, who presented to the diagnostic centre for FNAC (Fine needle aspiratrion cytology) procedure and CT scan. He had enlarged right cervical lymph nodes which were increasing in size progressively from last 6 months. There was no history of smoking and alcohol intake. Patient had no family history of tuberculosis or similar history of lymphadenopathy.

CT Scan revealed mildly enhancing enlarged cervical lymphnodes at right level II, III, IV and V with few nodes forming conglomerated groups, largest group was measuring 4.4 x 2.4 cm at level II. It was suggestive of haematological or neoplastic etiology more likely than infective etiology and correlation with FNAC for further workup was suggested.

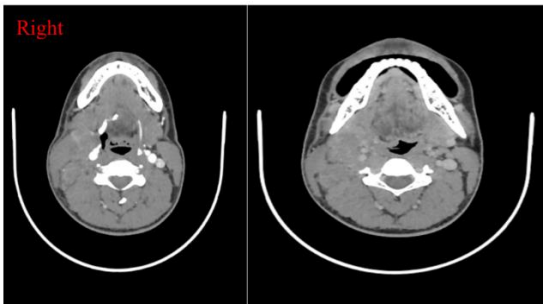

Figure 1: CT scan image: Showing conglomerated group of Right cervical lymphnodes.

1.1. Cytopathology

Patient underwent FNAC procedure. FNAC microscopy revealed a cellular smear with large atypical cells having large vesicular, indented, cleaved nuclei, central nucleoli and scanty cytoplasm. Few multinucleate giant cells with

*Corresponding author: Ranjana Hawaldar
Email: drshana.sampurna@gmail.com

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prominent nucleoli were seen. Frequent mitosis was present. Background showed lymphocytes, lymphoglandular bodies, RBC's and degenerated cells. No granuloma was appreciated in the smear studied.

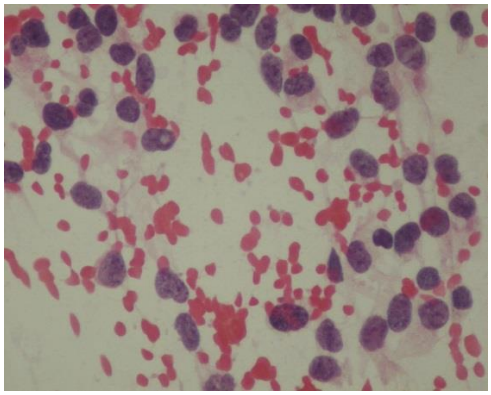

Figure 2: Giemsa stain (40x magnification): Large atypical cells with large vesicular, indented, cleaved nuclei, central nucleoli and scanty cytoplasm

Cytopathology reports were suggestive of Lymphoproliferative disorder. Excisional biospy and IHC was suggested for further evaluation.

1.2. Histopathological examination (Excisional biospy)

Patient underwent right cervical node excisonal biopsy by the general surgeon in a hospital set up. Biopsy was received for Histopathological examination.

Gross examination revealed a soft tissue mass measuring 1 cm in diameter grey white in color.

Microcopic examination revealed section showing lymph node with partially effaced architecture, dilated sinusoids, subcapsular sinuses containing round cells in small nests and loose clusters. Cells showed high N:C ratio, hyperchromatic nuclei and scanty cytoplasm. Few tumor giant cells were seen. Rest of lymph node showed normal morphology. Features were favouring two different diagnosis i.e. Metastatic round cell tumour and poorly differentiated carcinoma.

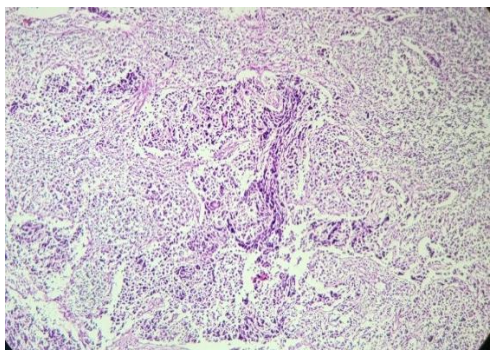

Figure 3: H&E stain (4x magnification): Image reveals lymphoid tissue showing dilated sinuses containing histiocytes and round cells with high N:C ratio, hyperchromatic nuclei and scanty cytoplasm

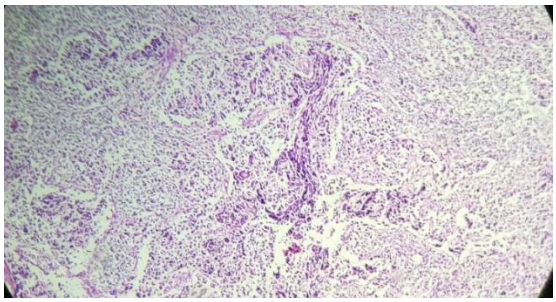

Figure 4: H&E stain (4x magnification): Dilated sinusoids with round cells

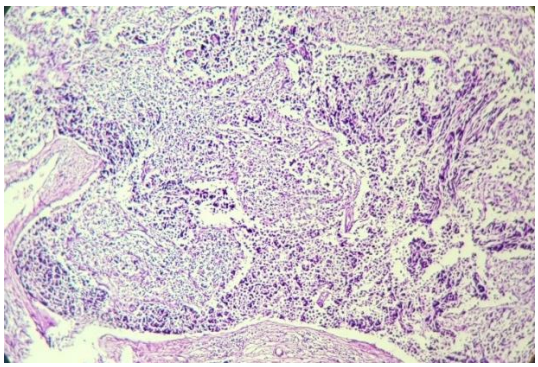

Figure 5: H&E stain (10x magnification): Dilated sinusoids and subcapsular sinuses containing round cells in small nests and loose clusters

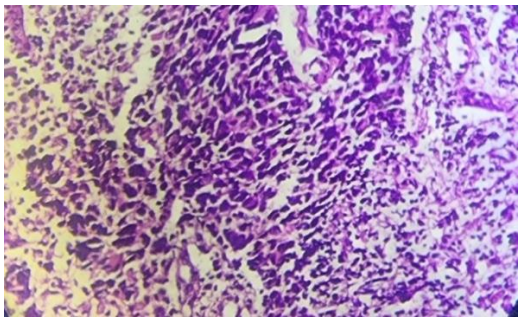

Figure 6: H&E stain (40 x magnification): Image reveals large round cells with hyperchromatic nuclei and eosinophilic cytoplasm

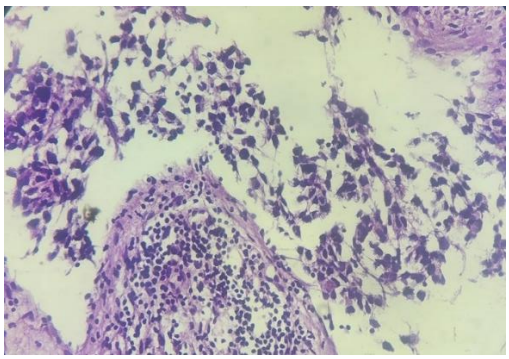

Figure 7: H&E stain (40 x magnification): Image reveals large round cells with hyperchromatic nuclei and an eosinophilic cytoplasm. Further Immunohistochemistry studies were conducted to further classify this lesion

1.3. Immunohistochemistry (IHC)

Immunohistochemistry (IHC) markers had a great role in further classifying and giving a definitive diagnosis of Alveolar Rhabdomyosarcoma (ARMS). Immunohistochemistry (IHC) relies on detecting positive staining for myogenic markers like desmin, myogenin, and MyoD1, along with negative staining for epithelial markers. Neoplastic cells were positive for myogenin and desmin.

Desmin: Desmin is a sensitive indicator of myogenic differentiation, especially in undifferentiated rhabdomyosarcomas.

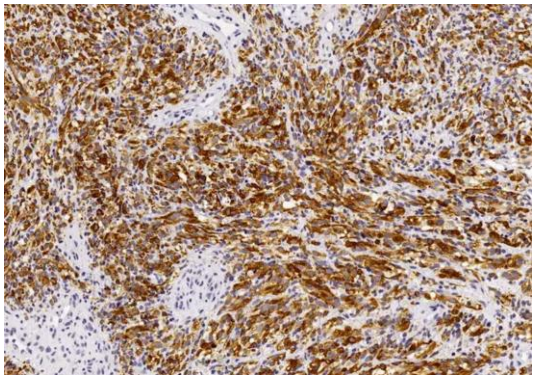

Figure 8: Desmin positivity (10 x magnification)

CD99: It is a cell surface antigen. It is typically expressed in a cytoplasmic pattern in rhabdomyosarcomas (RMS), while a membranous pattern is characteristic of Ewing sarcoma (ES).Therefore it rule out Ewing sarcoma.

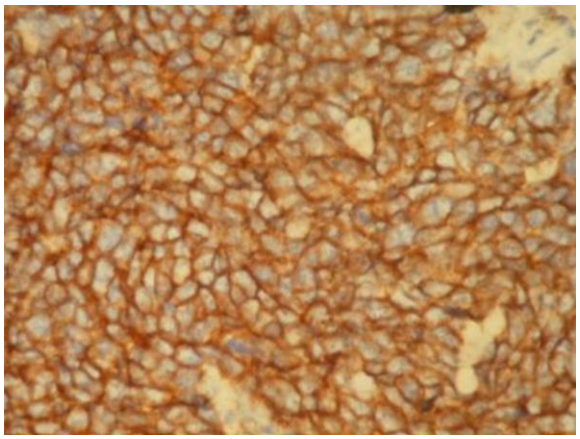

Figure 9: CD99 positivity (40 x magnification)

Myogenin: Alveolar rhabdomyosarcomas tend to exhibit stronger and more widespread myogenin staining compared to embryonal rhabdomyosarcomas. It is helpful in distinguishing alveolar rhabdomyosarcoma (ARMS) from embryonal rhabdomyosarcoma (ERMS).

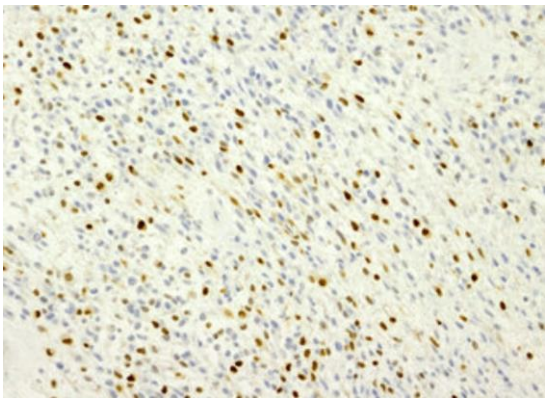

Figure 10: Myogenin (10 x magnification) -Focal positive

MyoD1: It is a myogenic regulatory protein, is commonly expressed in rhabdomyosarcoma (RMS) and is used as a diagnostic and prognostic marker.

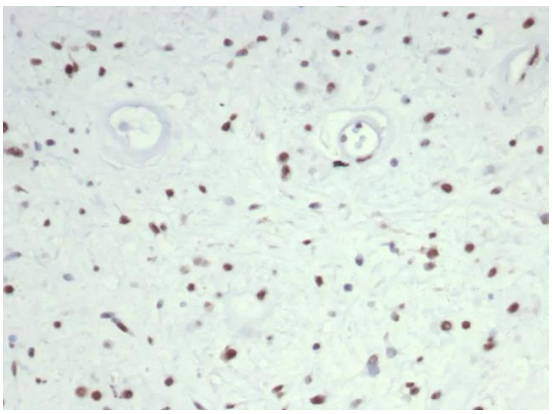

Figure 11: MyoD1 (10 magnification)-Focal positive

CD68 and HLADR - Highlights the admixed histiocytes
Other negative IHC markers
SMA, H caldesmin, CD31, P63, Pan CK-Negative
CK5/6, HMB 45, Melan A, SOX10, PAX 8- Negative
STAT 6, Synaptophysin, GFAP, EMA, SALL4- Negative
LCA, CD30, CD20, CD4- Negative
MPO, CD1a, CD23, TDT- Negative
Lysozyme, S100 - Negative

Table 1: IHC marker stain

IHC marker stain	Result	Application
Desmin	Positive	Myogenic differentiation
CD99	Positive	Cytoplasmic expression, cell surface marker
Myogenin	Focal positive	Specific to rhabdomyoblasts
CD 68	Positive	Identifying cells of the monocyte/macrophage lineage
HLADR	Positive	Expressed by antigen-presenting cells (APCs)
SMA	Negative	Identify smooth muscle tumours
H caldesmin	Negative	Specific to smooth muscle

CD31	Negative	Marker for endothelial cells
P63	Negative	Specific for Embryonal Rhabdomyosarcoma
Pan CK	Negative	Detection of Epithelial Origin Tumors
CK5/6	Negative	Specific for mesothelioma
HMB 45	Negative	Marker for melanoma
Melan A	Negative	Marker for melanoma
SOX10	Negative	Specific for melanocytic and Schwannian tumors
PAX 8	Negative	Specific for epithelial ovarian cancer
STAT 6	Negative	Specific for solitary fibrous tumor
Synaptophysin	Negative	Specific marker for neuronal differentiation
GFAP	Negative	Specific for astrocytes
EMA	Negative	Identify epithelial cells
SALL4	Negative	Expressed in germ cell tumor
LCA	Negative	Differentiated hematopoietic neoplasms and non-hematopoietic tumors
CD30	Negative	Key marker for Hodgkin lymphoma
CD20	Negative	Specific for B-cell lymphoma or leukemia
CD4	Negative	Surface marker for T helper cells
MPO	Negative	Specific marker for myeloid lineage
CD1a	Negative	Specific for dendritic cells
CD23	Negative	
TDT	Negative	
Lysozyme	Negative	
S100	Negative	

3. Discussion

Lymphadenopathic form of alveolar rhabdomyosarcoma shows lymph node involvement as first clinical manifestation in absence of recognizable primary tumor. There are few studies reporting this form of alveolar RMS.[8,9] Alveolar rhabdomyosarcoma can present with variable clinical and morphological presentation, may mimic lymphomas, leukemias and systemic metastatic disease with an unknown neoplasm.[9]

In 1958, Horn et al. introduced a widely accepted classification system that included embryonal, botryoid (a subtype of embryonal), alveolar,[3] and pleomorphic types of Rhabdomyosarcoma.[4] The World Health Organization (WHO) further categorizes this tumor into embroyonal, alveolar, pleomorphic and spindle cell/ sclerosing rhabdomyosarcoma.[5]

Immunohistochemistry staining with muscle related antigen (desmin, myogenin) is necessary to confirm the myogenic nature and confirm sarcomeric differentiation. Demonstration of positive Myogenin in a significant proportion of tumor cells, highlights the specificity of rhabdomyosarcoma, it is expressed in a particular strong and diffuse way in the alveolar type. Also desmin is an intermediate filament that is specific indicator for muscle differentiation.

Alveolar rhabdomyosarcoma (ARMS) should be differentiated from other small round blue cell tumors (SRBC) and soft tissue sarcomas. These are the differential which should be kept in mind while ruling out rhabdomyosacoma i. e. Ewing sarcoma, neuroblastoma, poorly differentiated synovial sarcoma, and other sarcomas like liposarcoma and osteosarcoma. Genetic testing can be helpful in confirming the diagnosis.

The standard treatment for adults with soft tissue sarcoma is based on surgery, complemented by radiotherapy, as Alveolar rhabdomyosarcoma are radiosensitive. To date, some benefit resulting from adjuvant chemotherapy has been demonstrated in adults with soft tissue sarcoma.[7]

Prognostic markers for Rhabdomyosarcoma are patient age, tumor size, tumor invasion, metastasis, regional lymph node involvement and tumor's response to chemotherapy. Patients who have been diagnosed with ARMS often have poor outcomes. The four-year survival rate without remission for local ARMS tumors is 65 percent, while the four-year survival rate with metastatic ARMS is only 15 percent.[10] Other variables affect the survival rate, such as primary tumor site, size of primary tumor, amount of local invasion, number of distal lymph nodes spread, and whether metastasis has occurred.[6,10]

4. Conclusion

Diagnosis of Alveolar rhabdomyosarcoma in a lymph node presents a challenge due to its uncommon occurrence. For diagnosis of Alveolar rhabdomyosarcoma (ARMS) cytopathology and histopathology is not always sufficient for an unequivocal diagnosis, necessitating ancillary studies, including immunohistochemistry (IHC).

5. Source of Funding

None.

6. Conflict of Interest

None

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