

Cytological diagnosis of metastatic deposits of alveolar rhabdomyosarcoma in ascitic fluid: A rare case report

Meera P.P., Bhulaxmi P., Nikhil S. Ghadyalpatil

ABSTRACT

Introduction: Rhabdomyosarcoma is an uncommon tumor seen in young adolescents and adults that metastasizes to the peritoneum. **Case Report:** Here we report a case of 18-year-old female who had alveolar rhabdomyosarcoma of nose and was treated with chemoradiotherapy. After one year, she presented with ascites. Ascitic fluid cytology revealed deposits of rhabdomyosarcoma. Immunohistochemistry done to support the diagnosis. This case report highlights the cytological features of rhabdomyosarcoma cells, which were large pleomorphic with high nuclear cytoplasmic ratio, coarse chromatin and scant cytoplasm. A small population of cells was almost the size of mesothelial cells, which could be mistaken for a reactive mesothelial cells. **Conclusion:** Awareness of rare metastases of rhabdomyosarcoma in peritoneum and recognition of the cytological features of these malignant cells supported by specific immunostains helps in the diagnosis.

Keywords: Cytological features, Immunohistochemistry, Myogenin, Rhabdomyosarcoma

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Received: 13 July 2016
Received: 15 September 2016
Published: 26 October 2016

How to cite this article

Meera PP, Bhulaxmi P, Ghadyalpatil NS. Cytological diagnosis of metastatic deposits of alveolar rhabdomyosarcoma in ascitic fluid: A rare case report. *Oncocytology* 2016;6:8–11.

Article ID: 100013OCYMP2016

doi:10.5348/OCY-2016-13-CR-3

INTRODUCTION

Rhabdomyosarcoma is one of the most aggressive soft tissue sarcoma that arises from the primitive striated muscle cells and is most commonly seen in the head and neck region [1]. Metastases develop during the course of the disease and are present at the time of diagnosis in about 20% of cases. Lung is the most common site of metastases in these tumors [2]. Here we report a rare case of alveolar rhabdomyosarcoma nose, who on posttreatment presented with ascites and ascitic fluid cytology revealed metastatic tumor deposits of rhabdomyosarcoma.

CASE REPORT

Case history

An 18-year-old female patient presented with abdominal discomfort and ascites paracentesis was performed and ascitic fluid was sent for biochemical analysis and cytological evaluation. Laboratory evaluation of ascitic fluid showed sugar 75 mg/dl, protein 4.8 g/dl, albumin 2.8 g/dl and LDH 7178 IU/ml. Ultrasonography of abdomen showed diffuse thickening

of omentum, hypoechoic lesion in peritoneum in the left hypochondrium measuring 90x46 mm along with moderate ascites. Two years earlier, the patient had presented with nasal polyp which was excised and on histology showed features of alveolar rhabdomyosarcoma. Patient was treated with chemoradiotherapy.

Past history

The patient had presented with a polyp in the middle turbinate for which polypectomy was done and sent for histopathological examination. It was reported as embryonal/ solid variant of alveolar rhabdomyosarcoma after immunohistochemical studies were done. Tumor cells were positive for Desmin, and negative for LCA, CK, CD99 and Synaptophysin. Patient also underwent positron emission topography scan for metastatic workup post-polypectomy which did not reveal any evidence of metabolically active disease elsewhere in the body. The patient received intergroup rhabdomyosarcoma study protocol IV (IRS-IV) which included vincristine, actinomycin D, cyclophosphamide alternating with vincristine, ifosfamide and etoposide. The patient also received radiotherapy to primary site at week-9.

Cytological findings

Centrifuged smears were stained with Giemsa. Hematoxylin and Eosin and Papanicolaou stain. Cell block was also made. Cytosmears revealed large cells in a dyscohesive pattern with high nuclear cytoplasmic ratio, coarse chromatin, indented nuclear membrane

and scant cytoplasm. Brisk mitotic activity noted in the background. These cells were admixed with mesothelial cells. Multinucleated cells were also noted. Few cells had irregular nuclei with complex convolutions ‘embryo like’ and ‘nose like’ protrusion (Figure 1A–C). These large cells were almost 2–3 times the size of mesothelial cells with similar nuclear features as mentioned above. Cytoplasmic vacuolation was noted in some cells. Nucleolus was inconspicuous. Good numbers of apoptotic bodies were also noted in the background. Pinkish hue was noted in the cytoplasm of these cells in contrast to the dense basophilic cytoplasm of mesothelial cells. No nuclear molding was seen. No lymphoglandular bodies were noted in the background.

Cell block was stained with hematoxylin and eosin revealed neoplastic cells with high nuclear cytoplasmic ratio, coarse chromatin, inconspicuous nucleoli and scant cytoplasm. No strap cells or classical rhabdomyoblasts were seen (Figure 1D).

The cytological features of the peritoneal fluid were initially interpreted as suggestive of malignancy and immunohistochemistry was performed so as to confirm whether it was a recurrence of alveolar rhabdomyosarcoma or any other secondary malignancy (for example, posttreatment induced secondary lymphoma). The neoplastic cells were positive for desmin (Figure 1E), myogenin (Figure 1F) and negative for LCA, NSE, CD56, CD99 and CK. A diagnosis of metastatic deposits of rhabdomyosarcoma was given based on cytological and immunohistochemical findings.

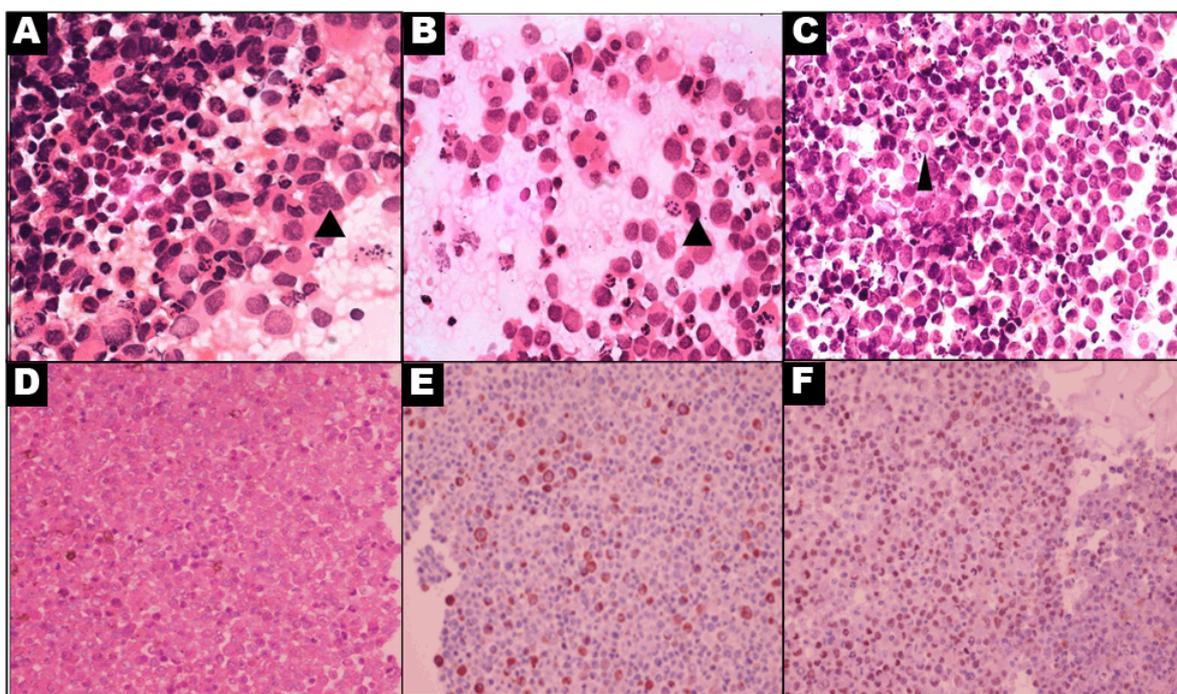


Figure 1: (A–C) Cytology smears of ascitic fluid. (Hematoxylin & Eosin stain all images, Original magnification x400) Neoplastic cells intermixed with reactive mesothelial cells (as depicted as arrow in figure C). “Embryo like” and multinucleated cell morphology of neoplastic cell is highlighted by an arrow in Figure A and B respectively. (D) Cell block of the same stained with Hematoxylin and Eosin, Original magnification x400), (E) Immunohistochemistry for desmin highlights cytoplasmic positivity in the neoplastic cells, and (F) Immunohistochemistry for myogenin shows strong nuclear positivity in the neoplastic cells, Original magnification x400).

DISCUSSION

Rhabdomyosarcoma is the most common soft tissue tumor of childhood, and is responsible for approximately one half of all soft tissue sarcoma in this age group. However they are rare, representing only 3–4% of pediatric cancer overall. Lung is the most frequent site of metastasis, other site of distant metastatic involvement include bone marrow (approximately 30%), bone (30%), omentum (ascites 16%), and pleura (13%) [3]. Rhabdomyosarcoma with primary in the perirectal tissue who clinically presented with ascites and was diagnosed on ascitic fluid cytology has been described in literature. Here is a rare case of rhabdomyosarcoma of nose who presented with ascites posttreatment and was diagnosed on ascitic fluid cytology as metastatic deposits of rhabdomyosarcoma which is very unusual of its kind.

The exfoliative cytology of rhabdomyosarcoma in body fluids has not been described in detail. However, fine needle aspiration cytology studies have shown that the cytomorphology of rhabdomyosarcoma is characterized by two main cell types, a predominantly primitive, small round cell with scant cytoplasm and a large cell with abundant cytoplasm, sometimes tadpole or ribbon-shaped. The primitive cells arranged in singles have a high nuclear cytoplasmic ratio and are small and lymphocyte like, with monotonous appearing nuclei which are round to polygonal with scant to moderate cytoplasm. The nuclear chromatin is fine granular and hyperchromatic while nucleoli are inconspicuous. Binucleated and multinucleated cells are seen often. The relative proportions of rhabdomyoblasts are seen more in embryonal rhabdomyosarcoma as compared to alveolar rhabdomyosarcoma. Binucleated cells, multinucleated cells, alveolar structures, cytological atypia are frequently encountered in alveolar rhabdomyosarcoma [4]. The presence of small undifferentiated cells in alveolar rhabdomyosarcoma makes the diagnosis more difficult and misinterpretation as hematolymphoid neoplasms are not uncommon, especially when metastatic to the bone marrow or lymph node [5].

Evaluation of effusion cytology is one of the most challenging areas in diagnostic cytopathology. A remarkably wide cytomorphological spectrum of reactive mesothelial cells overlap with various benign and malignant process. Cytological diagnosis of metastatic malignant tumor deposits in effusion cytology is made only after detection and confirmation of second cell population. The second cell population are 'non-inflammatory' and 'non-mesothelial'. Malignant cells shed into the fluids mimic reactive mesothelial cells including their characteristic 'window' which is generally seen in reactive mesothelial cells, further compounding to the diagnostic challenge in identifying them [6]. The case presented here had cytological features mimicking reactive mesothelial cells and also had nuclear convolutions mimicking a hematolymphoid malignancy.

The second foreign population of malignant cells in effusion is confirmed by immunohistochemical stains. Panel of immunostains are decided as per the suspected malignancy [4]. In this case, the panel of immunostains used based on the suspected second (malignant) cell population. In this case, the panel included desmin, LCA, myogenin, NSE, CD56, CD99 and CK.

Other malignant small round cell tumors considered in the differential diagnosis included desmoplastic small round cell tumor, Ewings sarcoma/primitive neuroectodermal tumor, lymphoma. Cytologically desmoplastic small round cell tumors have high nuclear cytoplasmic ratio, granular chromatin and also exhibits nuclear molding as a reminiscent of small cell carcinoma. Ewings/primitive neuroectodermal tumor exhibits dimorphic population of tumor cells with pale chromatin, 1–2 nucleoli and moderate vacuolated cytoplasm and darker staining cells with small nucleus with dense chromatin and narrow rim of cytoplasm. On immunohistochemistry tumor cells of desmoplastic small round cell tumors are immunoreactive for epithelial (keratin), mesenchymal (vimentin), myogenic (desmin) and neural markers (neuron specific enolase and CD56). Ewings sarcoma/primitive neuroectodermal tumor are immunoreactive for CD99 and neuron specific enolase and are negative for cytokeratin, desmin and leukocyte common antigen (LCA). Desmin is expressed by reactive mesothelial cells also [7–9].

Myogenin is a specific marker for rhabdomyosarcoma, as it is not expressed by mesothelial cells or other small round cell tumors that may also be encountered in effusion cytology, and have to be considered in the differential diagnosis of rhabdomyosarcoma, including desmoplastic small round cell tumor, Ewings sarcoma/primitive neuroectodermal tumor and other sarcoma, lymphomas and neuroblastoma and neuroendocrine carcinoma [10–12].

CONCLUSION

We present an unusual rare case of rhabdomyosarcoma of nose presenting with metastatic deposits in the peritoneum, and was diagnosed on ascitic fluid cytology. Cytology revealed predominantly large pleomorphic cells, which were larger than the mesothelial cells with high nuclear cytoplasmic ratio and coarser chromatin and admixed with small cells almost the size of mesothelial cells. Awareness of the rare metastases of rhabdomyosarcoma in the peritoneum and familiarity of the cytological appearance of these neoplastic cells supported by specific immunostains helps in the diagnosis.

Author Contributions

Meera P.P. – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Bhulaxmi P. – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Nikhil Ghadyal Patil – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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