Pleural effusion in a patient with Ewing sarcoma

Diane Frankel, Elise Kaspi, Corinne Bouvier, Patrice Roll

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1 | CASE HISTORY

A 25-year-old male was diagnosed with a Ewing sarcoma of the upper extremity involving the left tibia and fibula, with metastases in the left and right lungs. He was treated with radiotherapy and chemotherapy (carboplatin-etoposide).

Two years later, he presented with bilateral pleural effusions. Pleural fluid was aspirated and sent to the laboratory for cytological analysis (see images in Figure 1).

2. | MORPHOLOGY QUIZ

1. Based on the images provided (Figure 1), the pleural effusion contains:

A Macrophages
B Numerous mesothelial cells
C Eosinophils
D Dyskaryotic cells suspicious for malignancy

2. Based on pleural cytology and immunocytochemistry (Figures 1 and 2), what is the most likely diagnosis?

A Reactive mesothelial cells
B Metastatic non-small cell carcinoma
C Metastatic Ewing sarcoma
D Reactive hematopoietic cells

3. Which of the following antibodies is useful for Ewing sarcoma diagnosis using immunocytochemistry, and what is the staining pattern expected?

A Cytoplasmic expression of CD99
B Membranous expression of CD99
C Cytoplasmic expression of desmin
D Cytoplasmic expression of CD45

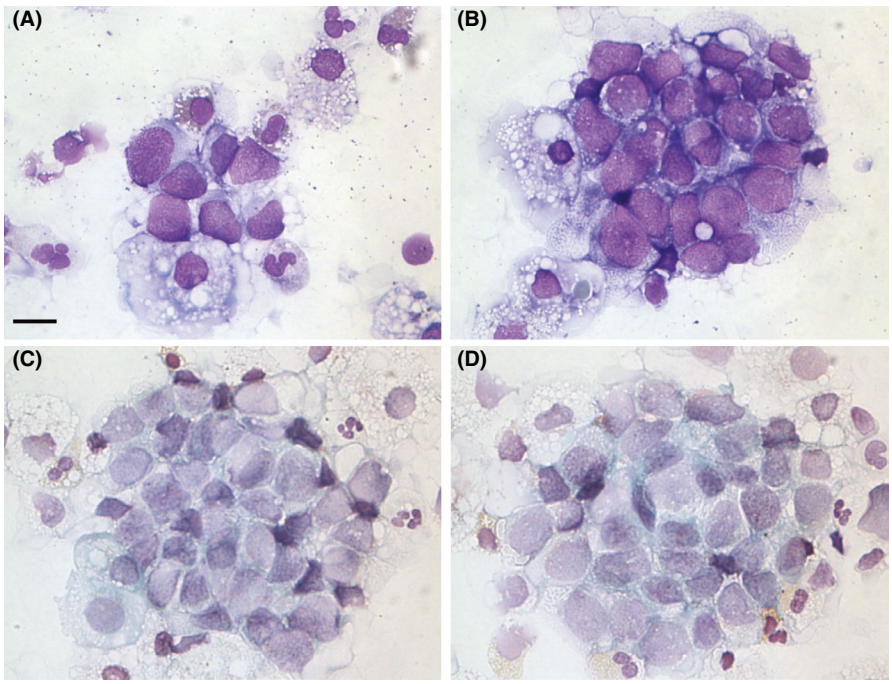

FIGURE 1 Cytology of pleural effusion (A, B: May-Grünwald-Giemsa stain, 40×; C, D: Papanicolaou stain, 40×; scale bar: 10 μm)

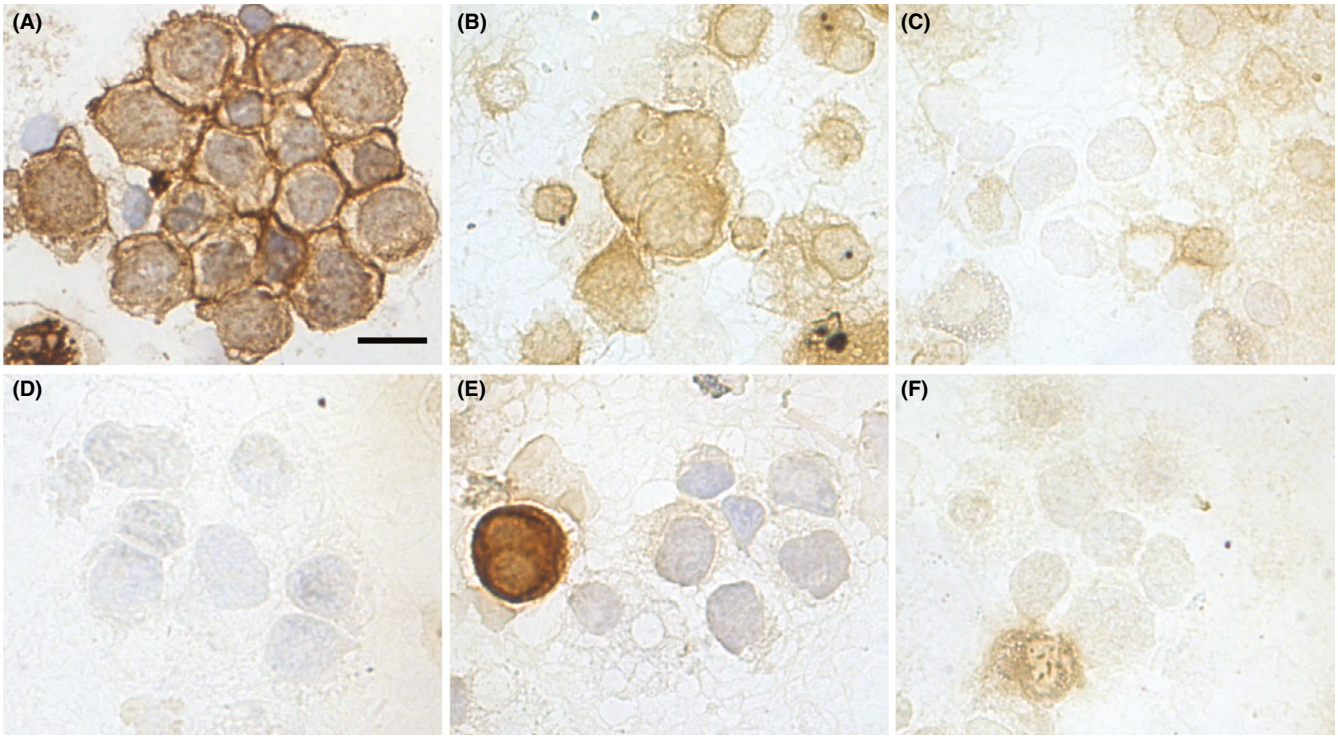

FIGURE 2 Immunocytochemistry (peroxidase staining, 40×) using antibodies against (A) CD99 (clone 12E7), (B) vimentin (clone V9), (C) CD45 (clone Mob040), (D) EpCAM (clone MOC31), (E) cytokeratins (clones AE1/AE3/PCK26). (F) Mouse IgG protein was used as isotypic control (scale bar: 10 μm)

4. What is the most common genetic abnormality seen in this tumour?

A *KRAS* mutation
B t(2;5)(p23;q35)
C t(11;22)(q24;q12)
D *ALK* rearrangement

3 | DISCUSSION

Ewing sarcoma is the second most common malignant bone tumour in children and young adults. The principal location is the diaphysis of long bones (pelvis, distal femur, proximal tibia, femoral diaphysis, or proximal humerus).[1] Twenty percent of patients present with metastases at the time of diagnosis, and lungs are one of the preferential metastasis sites, as are bone, bone marrow and lymph nodes. A metastatic pleural effusion is much less frequent since it is found in less than 10% of cases.

In the case presented in this article, the clinical context helps to establish the diagnosis. Without any context and based on the cytological examination, the presence of intermediate-sized cell clusters is indicative of a non-haematopoietic pathology. Immunocytochemistry must include epithelial, melanoma, sarcoma, and mesothelioma markers. The membranous expression of the CD99 associated with the negative expression of epithelial and melanoma markers confirms the diagnosis. A genetic study must also be carried out, and the identification of the characteristic t(11;22) with EWS/FLI1 fusion gene confirms the diagnosis.

The present report, in addition to documenting a rare case, highlights the pivotal role of pleural cytological examination in the diagnosis of such a case.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

AUTHOR CONTRIBUTIONS

DF, EK and CB: Data collection; DF: writing the manuscript; EK, PR and CB: reviewing the manuscript.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

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ANSWERS TO MORPHOLOGY QUIZ

Question 1

Answer: A, C and D.

Question 2

Answer: C.

Question 3

Answer: B.

Question 4

Answer: C.