FNA DIAGNOSIS OF SIGNET RING CELL LYMPHOMA: CASE SERIES AND LITERATURE REVIEW

Anne Tu1, Chris Van Vliet1, Sue Sparrow1, Ming Chai1, and Gregory Sterrett1.
1: PathWest Anatomical Pathology Services, Cytology Division, Perth WA

INTRODUCTION

Signet ring cell lymphoma (SRCL) is a rare morphologic variant of non-Hodgkin lymphoma. This phenomenon has been described most commonly in follicular lymphoma but has also been described in B-cell lymphomas with plasmacytic differentiation and very rarely in T-cell lymphomas1-4. We retrospectively reviewed three fine needle aspiration (FNA) cases of lymphoma with signet ring features retrieved from our case files.

CLINICAL PRESENTATIONS

CASE 1: 54 year-old male presented with pulmonary embolism and deep vein thrombosis. CT chest, abdomen and pelvis showed an ill-defined opacity in the right upper lobe of the lung with associated pleural thickening, an enlarged lymph node in the right hilum and a retroperitoneal mass inferior to the head of pancreas with slightly enlarged nodes in the para-aortic region. EUS FNA of the peri-pancreatic mass was performed and material was triaged for flow cytometry.

CASE 2: 59 year-old male with known non-Hodgkin B-cell lymphoma. PET scan showed a new intensely active pleural based mass. Instead, material was collected for cell block and immunohistochemistry.

CASE 3: 67 year-old male presented with left inguinal lymphadenopathy. CT abdomen showed multiple enlarged nodes throughout the posterior mediastinal and inguinal regions along with large lobulated masses within the retroperitoneum and pelvis. US FNA and concurrent core biopsy of the left inguinal lymph node was performed and material was triaged for flow cytometry.

ANCILLARY TESTING

FLOW CYTOMETRY: Case 2 and 3 were triaged for flow cytometry and both showed a CD10-positive monoclonal B-cell population with immunoglobulin light chain restriction.

IMMUNOHISTOCHEMISTRY: Material from the case 1 was not triaged for flow cytometry at rapid onsite evaluation as non-haematolymphoid malignancy was suspected initially. Instead, material was collected for cell block and immunohistochemistry showed strong positive staining for CD45, CD20, CD10, Bcl2 and Bcl6 in the abnormal population of centrocytes and signet ring forms. Staining for epithelial, melanocytic and soft tissue markers were negative.

FLUORESCENCE IN-SITU HYBRIDISATION: An IGH-BCL2 dual fusion probe confirmed IGH-BCL2 translocation.

DISCUSSION

Signet ring change is rare in non-Hodgkin lymphoma. This phenomenon has been described most commonly in follicular lymphoma, which coincides with our experience. It has also been described in B-cell lymphomas with plasmacytic differentiation including diffuse large B-cell lymphoma, marginal zone lymphoma and lymphoplasmacytic lymphoma and very rarely in T-cell lymphomas1-4. In B-cell lymphomas, the signet ring appearance is due to cytoplasmic accumulation of immunoglobulin5-7, usually IgG as large clear vacuoles or IgM as eosinophilic globules. In T-cell lymphomas, these vacuoles are thought to be derived from multivesicular bodies.

SRCL do not differ from their non-signet ring counterparts1-4 in their clinical presentation (nodal or extranodal), behaviour, immunophenotype or cytogenetics. They may present a diagnostic challenge and the main pitfall is misdiagnosis of SRCL as other non-haematolymphoid entities that exhibit signet ring features1-4, in particular adenocarcinoma (as occurred in case 1) as well as melanoma, liposarcoma, squamous cell carcinoma, mesothelioma and benign histiocytic reactions.

TAKE HOME MESSAGE:

• Signet ring cell change is rare in non-Hodgkin lymphoma with most reported cases occurring in follicular lymphoma
• Signet ring cell lymphoma can mimic other non-haematolymphoid entities

REFERENCES