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Turkish Society of Hematology

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EHA-TSH Hematology Tutorial on Lymphoma

Tutored Clinical Case 1

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İzmir, Turkey
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Clinical history

- 29 year old female patient presented with an enlarged cervical lymph node.
- A course of antibiotics was administered without a change in the size of the lymphadenopathy.
- A fine needle aspiration performed from the mass was interpreted as consistent with reactive changes at another institution.



Clinical history

- An excisional biopsy of the lymph node was performed because the patient was concerned about her physical appearance.
- Pathological evaluation at another institution was considered as suspicious for lymphoma.
- The patient requested a second opinion at our Institution.



Clinical history

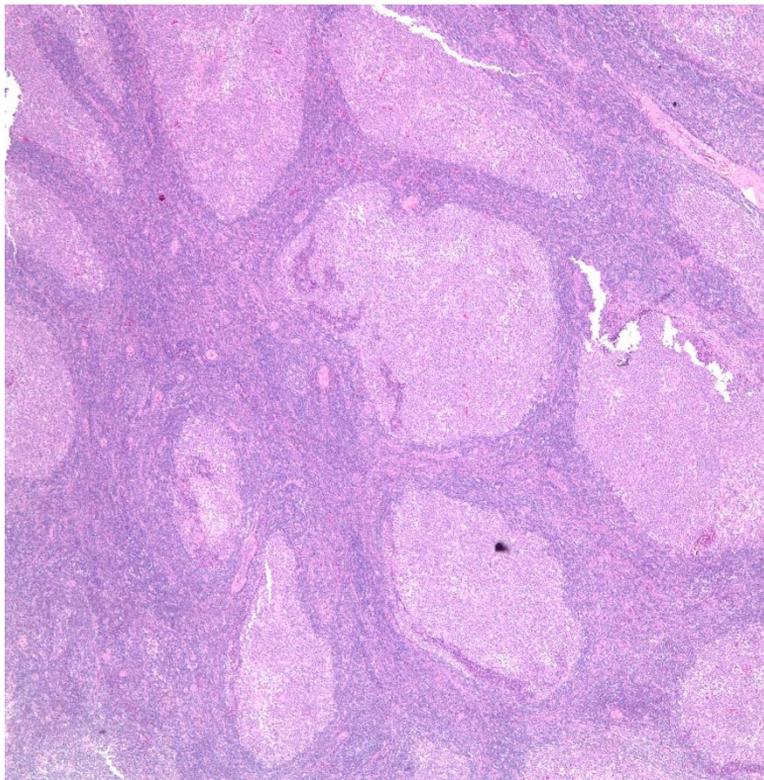
- Physical examination showed an incision scar for the submandibular lymph node. There was no additional lymphadenopathy or hepatosplenomegaly. There were also no B symptoms.
- Past medical history was not significant for any diseases.
- Radiological examination and laboratory results were all within normal limits.



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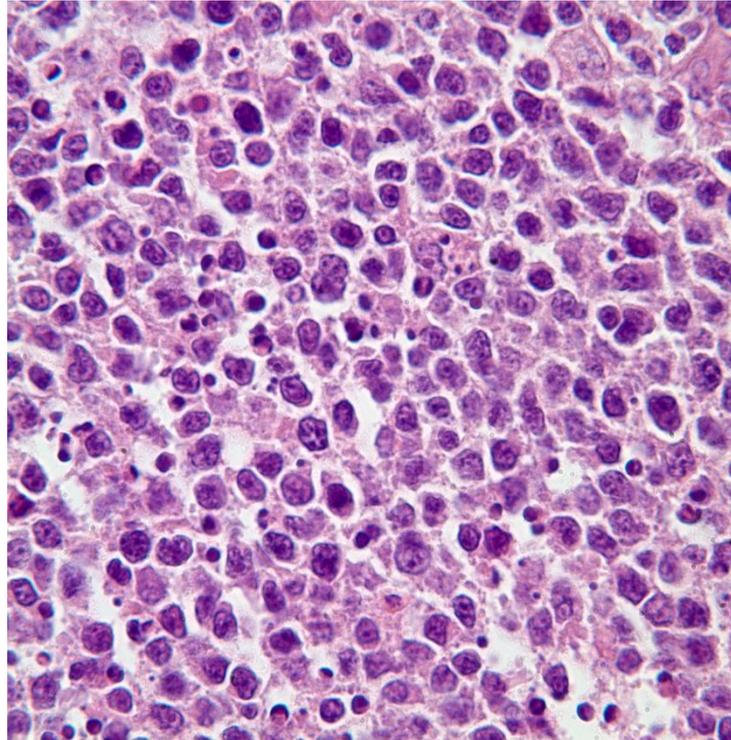
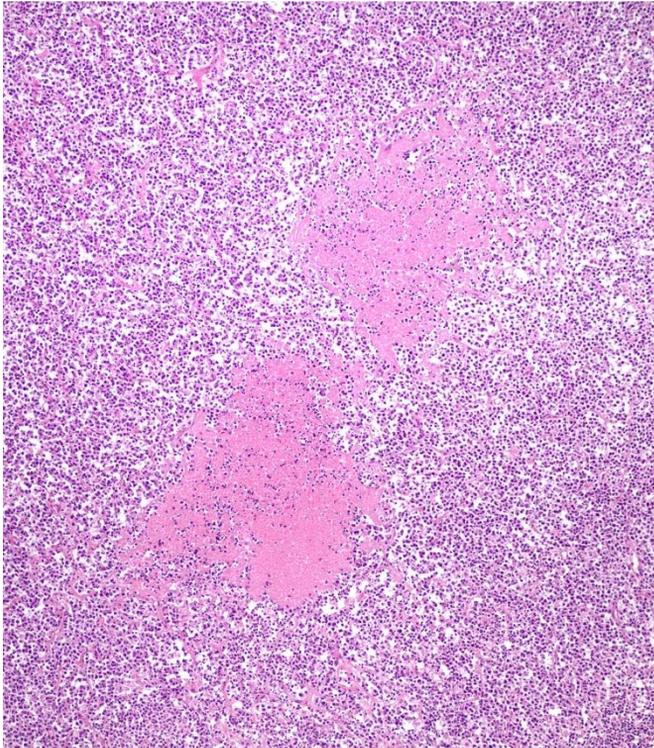
Lymph node biopsy





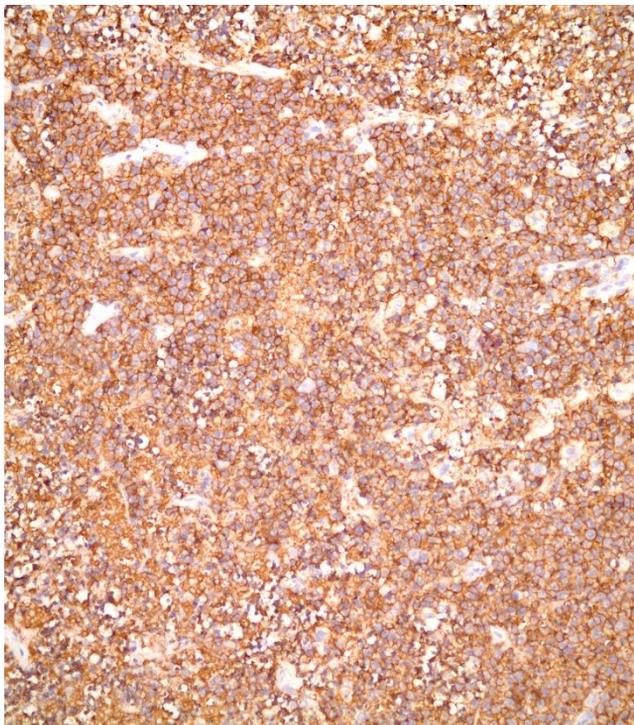
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Higher magnification

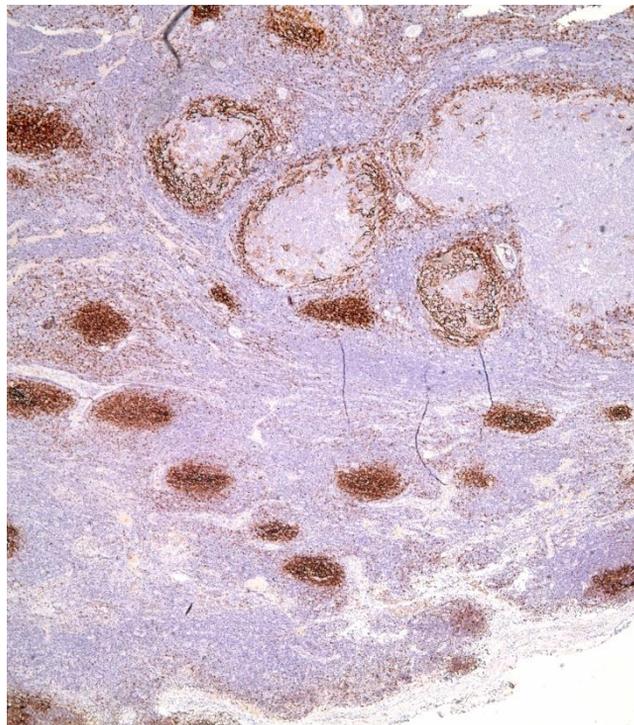




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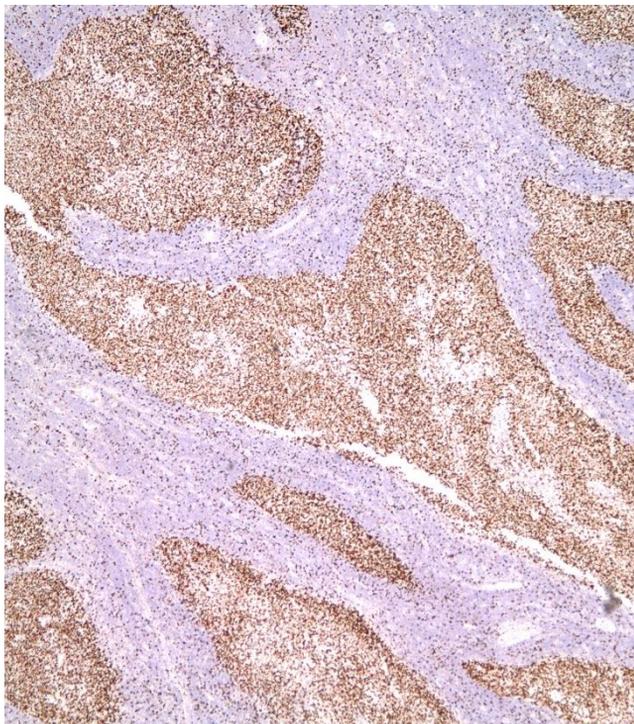
CD20



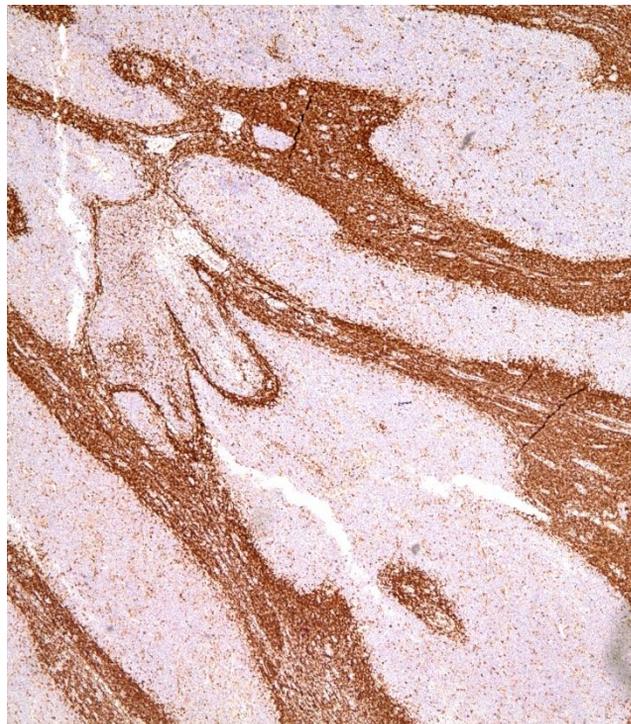
CD23



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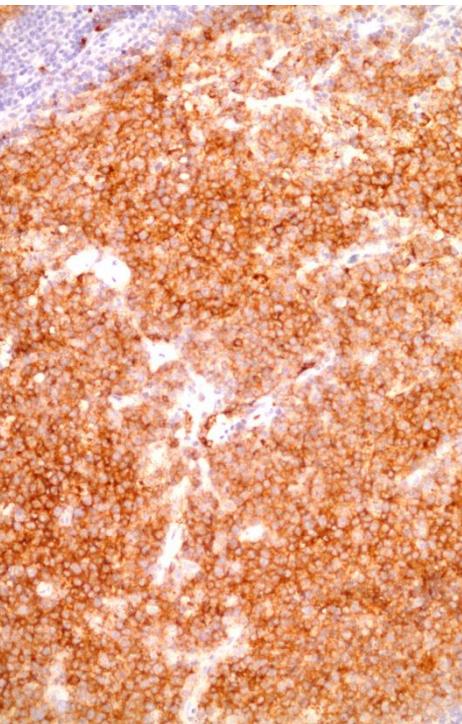
Ki-67



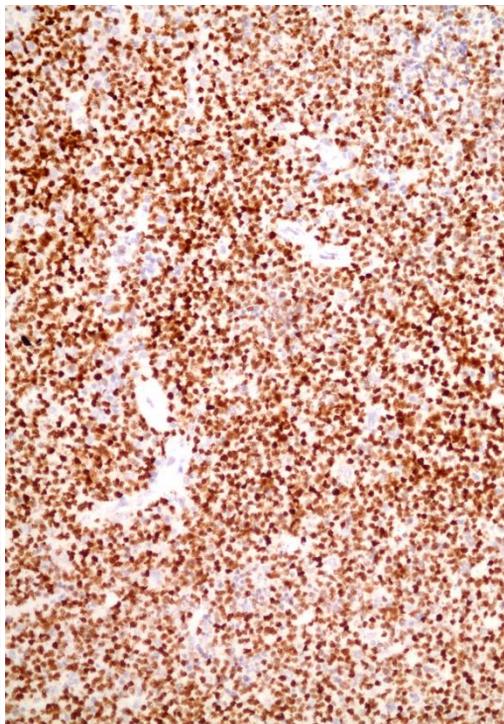
Bcl-2



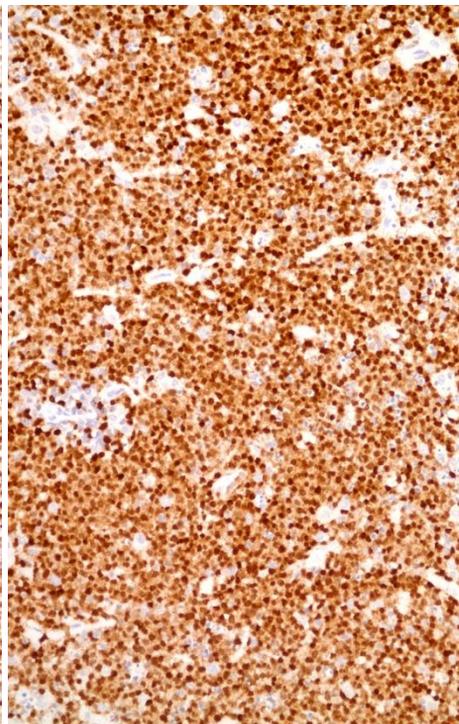
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CD10



Bcl-6



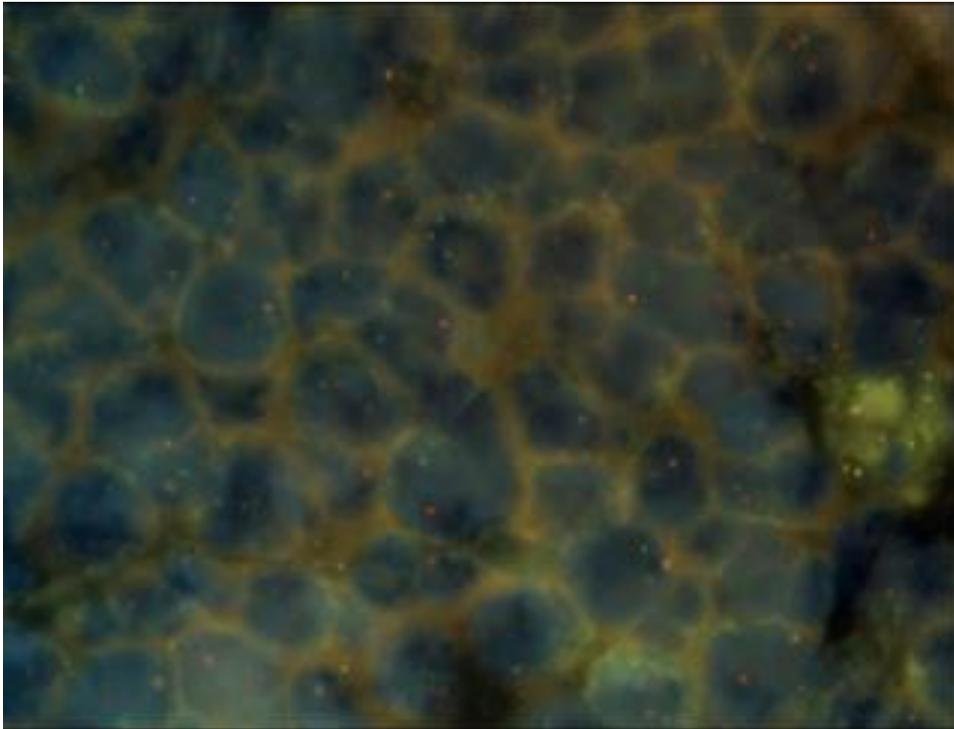
MUM-1



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IRF4/DUSP22 FISH Breakapart

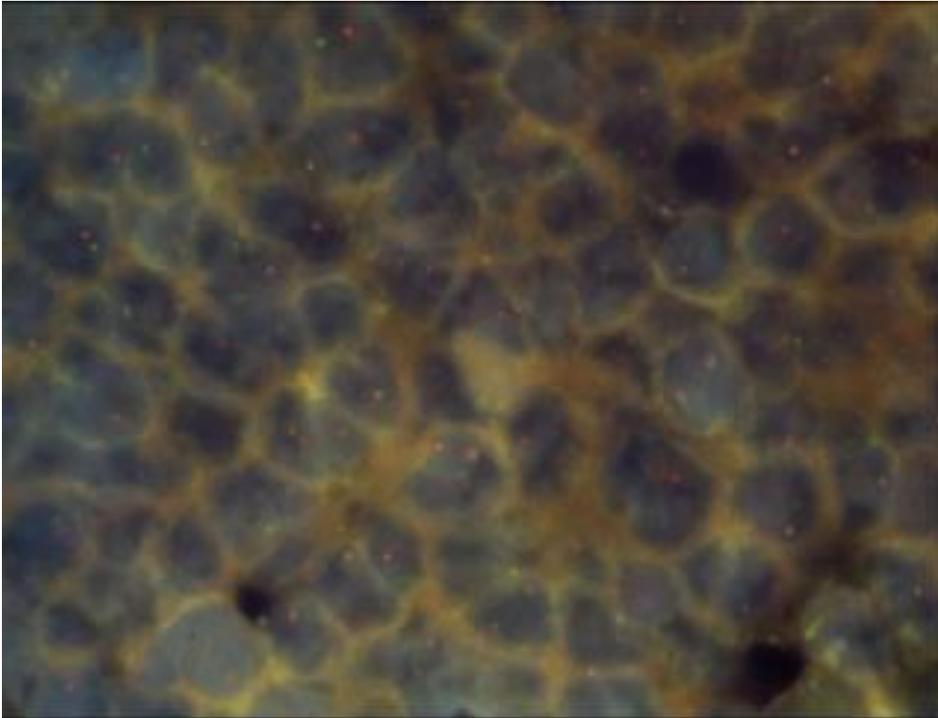




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IRF4/DUSP22 FISH Breakapart





Diagnosis

- Large B- cell lymphoma associated with *IRF4*-rearrangement (WHO-2016)
- Follicular lymphoma, grade 3B, nodular(%75) ve diffuse (%25) (WHO-2008)
- FISH: Normal pattern for *MYC*, *BCL-2* and *BCL-6* genes. Breakapart probe for *IRF4/DUSP22* showed evidence of translocation



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Additional Clinical Information

- No evidence of bone marrow involvement
- Stage I
- The patient did not receive any treatment
- No evidence of disease with follow-up of more than two years.



Large B-cell lymphoma with IRF4 rearrangement

- Rare type of lymphoma accounting for 0.05% of large B cell lymphomas.
- Most patients present with isolated enlarged lymph node of the head and neck region or involvement of the Waldeyer Ring. Involvement of GI tract is also reported



Large B-cell lymphoma with IRF4 rearrangement

- Primarily disease of children and young adults (median age 12; age range 4-79 years)
- Equal sex distribution
- Usually early stage
- Favourable outcome (5-year survival 100%)



Large B-cell lymphoma with *IRF4* rearrangement

- Cases can be classified as follicular lymphoma (pediatric-type). Distinction is important because of management issues
- Co-expression of CD10, BCL-6 and MUM-1 is an important clue. Such cases should be screened for *IRF4* rearrangement



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Reference

- Salaverria I *et al* (2011) Translocations activating IRF4 identify a subtype of germinal center-derived B-cell lymphoma affecting predominantly children and young adults. *Blood*, 118(1):139-47.