

Three cases of Epstein-Barr virus-positive plasmacytoma in immunocompetent patients: a diagnostic dilemma

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Introduction

- Plasmacytomas
 - solitary plasmacytoma of bone (SPB)
 - extraosseous/extramedullary plasmacytoma (EMP)
 - EBV-related plasmacytomas: rarely reported
- Plasmablastic lymphomas (PBL)
 - typically EBV positive
 - frequently in HIV⁺ or other immunocompromised individuals
- EBV-positive plasmacytomas (especially plasmacytomas with anaplasia characteristis) may morphologically and phenotypically resemble with PBL

3 Cases of EBV-positive Plasmacytoma

	age	gender	Immuno-deficiency history	site	symptoms	Radiologic findings	bone lesions
1	45	M	-	thoracic vertebra	backache, weakness of lower extremities, urination defecation difficulties	bursting fracture of thoracic vertebra	+
2	69	F	-	Left nasal cavity	Inhale with difficulty, nasal obstruction with mild rhinorrhagia	a mass in nasal cavity	-
3	26	M	-	inferior nasal concha	dry eyes	maxillary sinusitis	-

3 Cases of EBV-positive Plasmacytoma

HE

CD79a

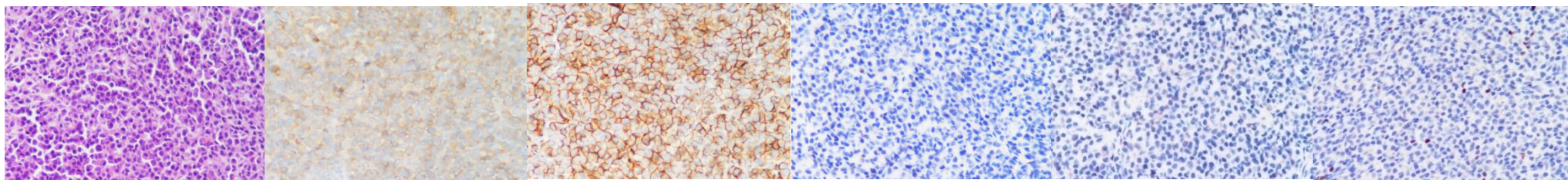
CD138

κ

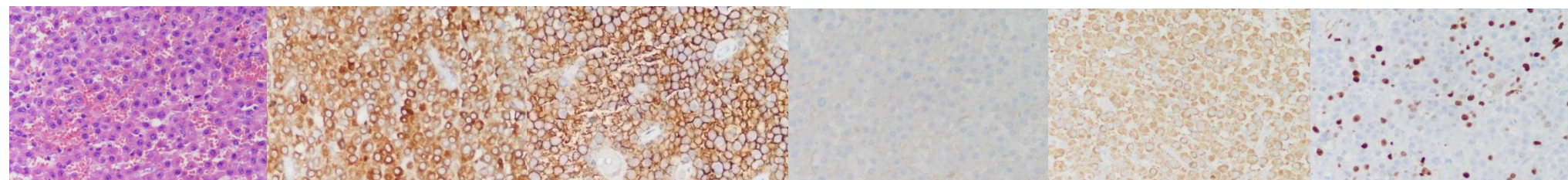
λ

Ki67

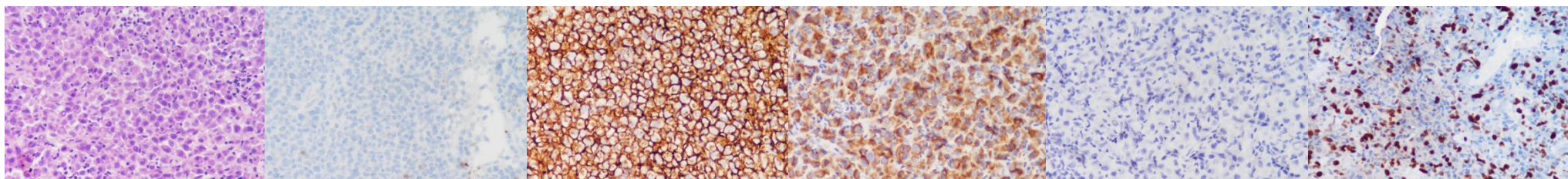
Case 1



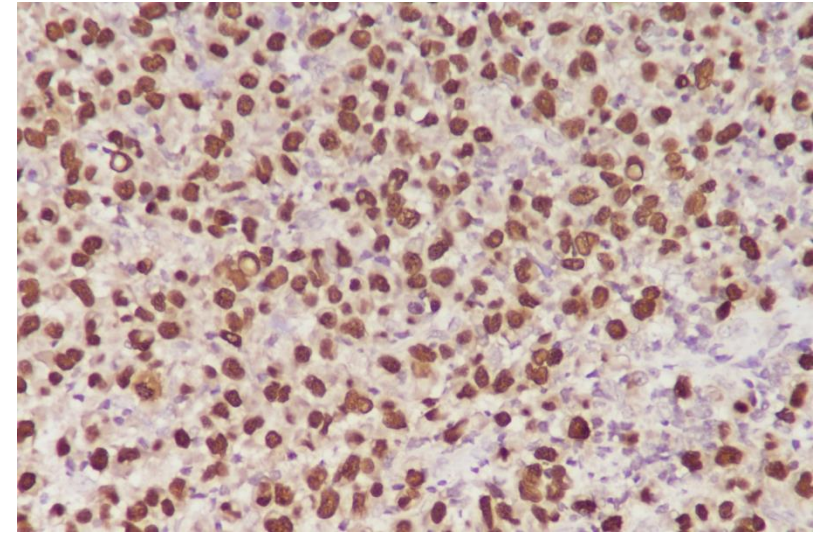
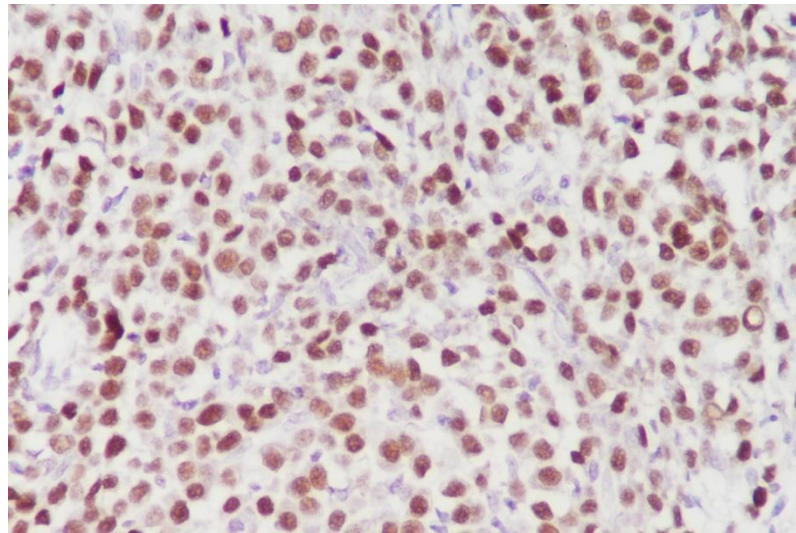
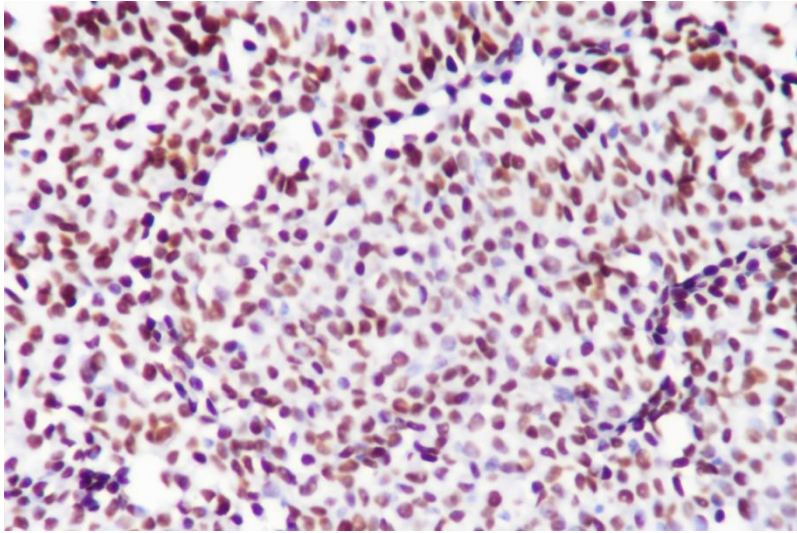
Case 2



Case 3



EBER for 3 Cases



3 Cases of EBV-positive Plasmacytoma

Case	EBER	CD20	CD79α	CD138	CD38	MUM-1	CD56	Cyclin D1	Light chain	Ki67
1	+	-	+	+	+	ND	-	-	κ(-), λ(-)	2%
2	+	ND	+	+	+	+	+	+	κ(-), λ(+)	20%
3	+	ND	-	+	+	-	ND	ND	κ(+), λ(-)	15%

3 Cases of EBV-positive Plasmacytoma

Case	IG rearrangement	FISH	BM	immunofixation electrophoresis
1	IGκ-VJ rearrangement	1q21,13q14 (RB1) , 13q14.3 (D13S319) , IGH	Plasma cells (1.5%)	Polyimmunoglobulin
2	ND	ND	-	NA
3	ND	ND	-	NA

3 Cases of EBV-positive Plasmacytoma

Case	therapy	Outcome (months)
1	Lenalidomide + dexamethasone	Alive (31)
2	Chemotherapy + radiotherapy	Alive (12)
3	radiotherapy	Alive (65)

Discussion

- Solitary plasmacytomas
 - monomorphic, well-differentiated/atypical neoplastic plasma cells
 - indistinguishable from PBL
 - bones: most commonly involved sites
 - Two types of solitary plasmacytoma: SPB & EMP
 - EMP
 - typically involve the upper respiratory tract of immunocompetent individuals
 - usually respond to radiation therapy or limited cycles of chemotherapy
- ***EBV-positive plasmacytoma in Immunocompetent patients (EPIC)***

Discussion

- PBL
 - clinically aggressive B-cell neoplasms
 - lack B-cell markers but express plasma cell markers
 - generally a **high Ki67 proliferative index (often >60%)**
 - often show **MYC abnormalities**
 - approximately **75-100%** of PBL are **positive for EBER??**
 - typically in **immunocompromised** patients
 - PBL in immunocompetent individuals have been described not frequently

Discussion

- EBV-positive plasmacytoma may morphologically and phenotypically mirror PBL, especially anaplastic plasmacytoma
- EBV infection status is **no longer enough** to help determine the type
- The morphology is always the most important.
- Other diagnostic clues include **relatively low Ki67 proliferative index (usually <20%), MYC aberrations** in plasmacytoma as well as **immunodeficiency history**.
- The genetics of plasmacytomas have not been extensively studied so far.

Discussion

- EBV⁺ plasmacytomas VS. EBV⁻plasmacytomas
 - do they perform distinctively in clinicopathological characteristics, genetic profiling, biological behaviors and prognosis?
 - Is there a possibility that they belong to different entities?
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- Further larger-scale multicenter studies are warranted.

Thank you for your attention!