

Supplementary data

Supplement to: Vogrig A, Péricart S, Pinto AL et al. Immunopathogenesis and proposed clinical score for identifying Kelch-like Protein-11 encephalitis. *Brain Communications* 2021

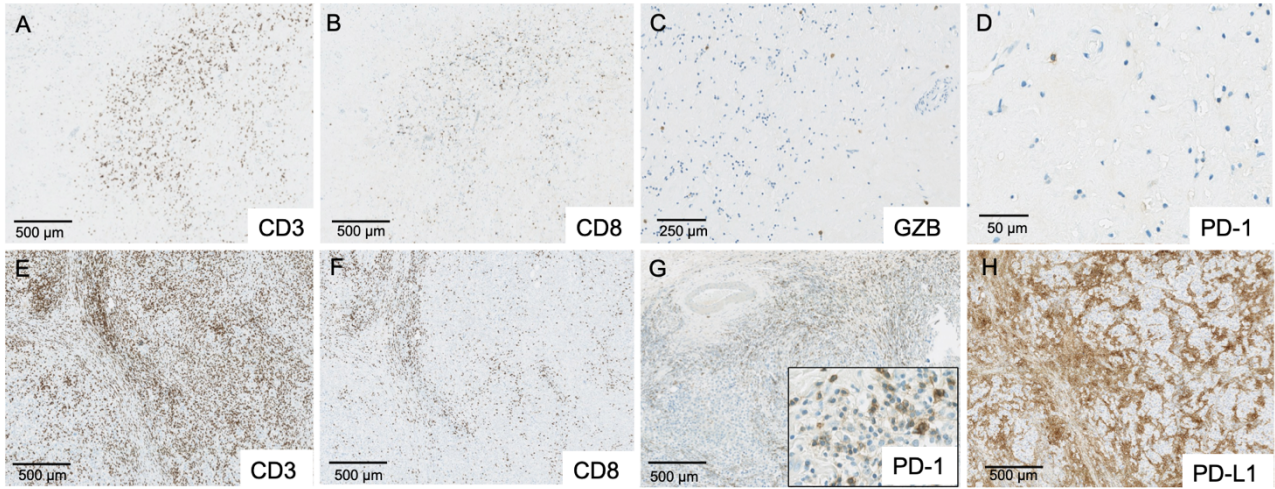
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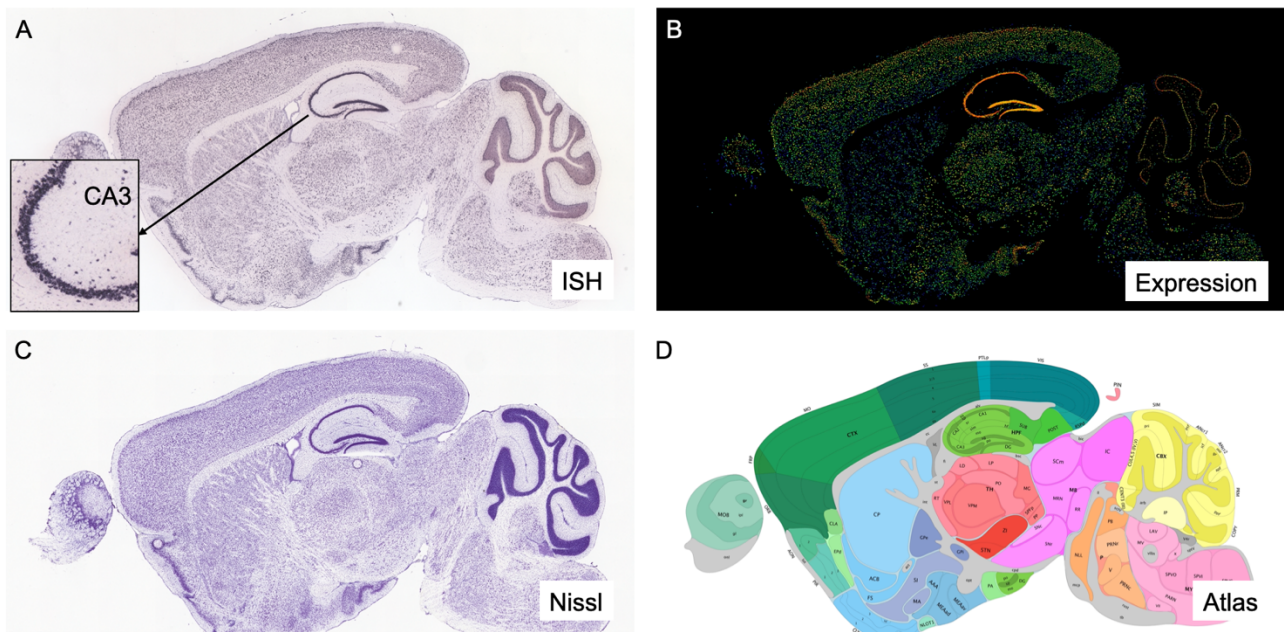
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Supplementary Figure 1: Immunohistochemical studies in primary testicular tumours from control patients without KLHL11-Abs



A-B: Immune profile in a «burned-out» testicular tumour (A: IHC anti-CD3 ($\times 100$), B: IHC anti-CD8 ($\times 100$), C: IHC anti-Granzyme B ($\times 150$), D: IHC anti-PD-1 ($\times 300$). E-H: Immune profile in a classical form of seminoma (E: IHC anti-CD3($\times 100$), F: IHC anti-CD8($\times 100$), G: IHC anti-PD-1($\times 100$, insert: $\times 300$), H: IHC anti-PD-L1 ($\times 100$).

Supplementary Figure 2: Expression of *KLHL* in the mouse brain



In situ hybridization (ISH) to localize the specific sequence of the *KLHL* gene using an RNA probe (A) and *KLHL* gene expression (B), as compared to Nissl staining (C), a classic nucleic staining method used on nervous tissue sections, and (D) a mouse brain atlas, shown to highlight the regions involved (hippocampus, including the CA3 region, as detailed in A, as well as the cerebellum).

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Supplementary Table: Neuropsychological profiles in patients with with KLHL11-Abs and complete cognitive evaluation

Patient N°	Cerebellar cognitive affective syndrome (CCAS)				Extra-CCAS cognitive impairment	Neuropsychological report (n°) and conclusions
	Executive dysfunction	Linguistic Processing deficits	Spatial cognition deficits	Affect dysregulation		
1	X			X		(1) Dysexecutive syndrome, behavioural alterations, attention deficits. Anterograde memory not affected (2) Cognitive profile stable 10 months later (3) Similar cognitive profile 9 months later. Marked verbal and motor perseverations. Less behavioural alterations but persistence of hyper-emotivity and low-frustration tolerance MMSE: 23/30, FAB: 7/18
4	X	X		X		(1) Dysexecutive syndrome with prominent deficits in planning, mental flexibility, and attention. Language production deficit. Difficulties in drawing and writing. Cognitive fatigability. Relatively preserved domains: working memory, episodic memory, visuo-spatial functions (2) Cognitive profile stable 5 months later
5	X	X	X	X	Hippocampal	(1) Massive deficit in verbal episodic memory, visuospatial exploration deficits, impaired attention. Anxiety-depressive disorder (MADR 20/60; Goldberg 8/9). MMSE 26/30 (2) Deficit in verbal episodic memory and global alteration of cognitive functions 34 months later. MMSE 25/30
6	X	X	X	X	Hippocampal	(1) Dysexecutive syndrome with deficits in mental flexibility and impaired inhibition. Working memory deficits. Impaired verbal and visual episodic memory. Anxiety
8		X	X	X	Hippocampal	(1) Deficits in episodic verbal memory (Grober & Buschke). Normal encoding (Rim), storage (RT1), and consolidation (RD). Deficits in nonverbal episodic memory. Normal flexibility (TMT-B), ability to inhibit cognitive interference (Stroop), and cognitive processing speed (TMT-A, Stroop). Deficits in visuospatial attention (Mesulam & Weintraub). Decreased verbal production and fluency, deficits in oral denomination (DO80), anxiety. Normal praxis abilities (Mahieux). MOCA 28/30
9	X	X		X	Hippocampal	(1) Dysexecutive syndrome with alteration in planning, mental flexibility. Agrammatism. Deficit of episodic memory (verbal and visual) and working memory. Apathy, abulia, anhedonia. MMSE: 18/30, BEC 96: 55/96, Clock drawing test: 5/7, FAB: 7/18, Praxis: symbolic gestures 3/5, action mimicking 10/10, abstract gestures 5/8

Abbreviations: BEC, Batterie d'évaluation cognitive (cognitive evaluation battery); CCAS, Cerebellar cognitive affective syndrome; FAB, frontal assessment battery; MADR, Montgomery-Åsberg Depression Rating Scale; MMSE, Mini Mental State Examination; RT1, Reaction Time test 1; TMT, Trail Making Test