Universitätsklinikum Erlangen

Addisonian crisis due to autoimmune adrenalitis in a 14 years old boy after haematopoietic stem cell transplantation (HSCT).

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Objective

To report on a primary adrenal insufficiency in a 14 years old boy from Albania.

The diagnosis was made 12.7 years after HSCT due to an Addisonian crisis.

Conclusions

- Primary adrenal insufficiency in children due to autoimmune adrenalitis is an uncommon condition in children after HSCT.
- We speculate that our patient has autoimmune polyglandular syndrome (APS) type 2 with no coherence to the HSCT.

Background

Endocrine complications after HSCT are common and mainly related to the myeloablative therapy associated with total body irradiation or immunosuppression.

These complications can develop over a long period.

Secondary adrenal insufficiency due to suppression of the hypothalamic-pituitary-adrenal axis mainly depends on the duration and cumulative dose of corticosteroid treatment after HSCT.

To the best of our knowledge, we found only two reports on primary adrenal insufficiency after HSCT in the literature.

Primary adrenal insufficiency (PAI) after HSCT in the literature

Age, sex	Primary Dx.	Myeloablative regimen	Dx of PAI	Cortisol nmol/L	ACTH pg/ml	Aetiology
9 years, girl (1)	Myelo-dysplastic syndrome	Busulfan, Cyclophos-phamide	23 mo. after HSCT	205 Low-dose ACTH test: no stim.	595	unknown
11 years, boy (2)	Sickle cell anaemia	Busulfan, Cyclophos-phamide, antithymo-cyte globulin	4 mo. after HSCT	55 High-dose ACTH test: no stim.	normal (analytical problem)	APS type 2

Case report

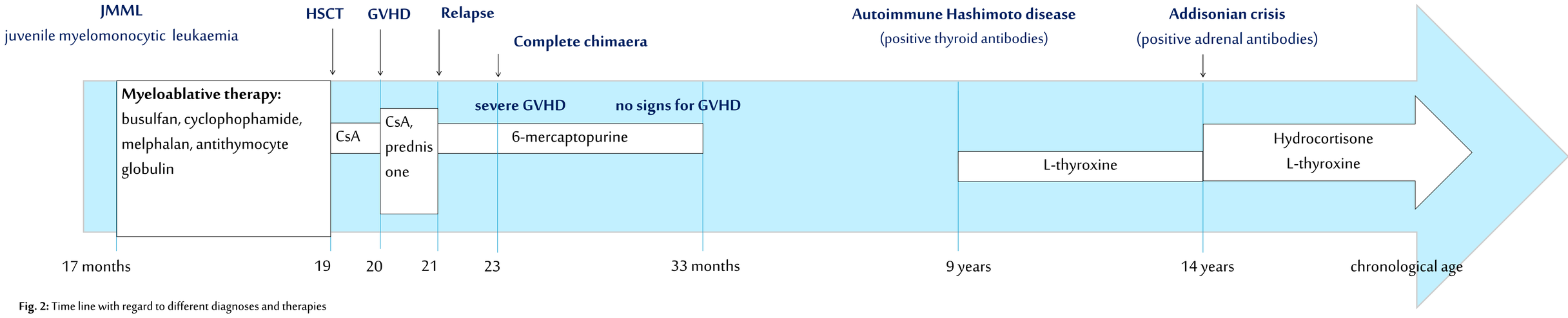

Fig. 2: Time line with regard to different diagnoses and therapies

	Last outpatient visit	Addisonian crisis (two weeks later)
Clinical parameters	• good general condition • soccer player • RR 122/76 mmHg • HR 73/min	• reduced general condition • nausea • weight loss 1.3 kg • RR 99/61 mmHg • HR 116/min
Auxology and Tanner stage	• Height 165.5 cm (-0.37 SDS) • Weight 46.8 • BMI 17.1 kg/m² (-0.9 SDS) • Tanner pH 3 • testes size (ml) 8/8	• Height 165.5 cm (-0.37 SDS) • Weight 45.5 kg • BMI 16.6 kg/m² (-1.3 SDS) • Tanner pH 3 • testes size (ml) 8/8
Electrolytes (serum; mmol/l)	• sodium 133 (132-141) • potassium 4.2 (3.3 - 4.6)	• sodium 114 (132-141) • potassium 6.0 (3.3 - 4.6)
Cortisol (nmol/l; >138) 8 – 10 am	151	264
ACTH (plasma pg/ml; 7.2-63)	Nd	625
Renin (serum; pg/ml; 8.8-76)	Nd	2550
Antibodies	• Tg > 2000 U/ml (< 60) • TPO 1941 U/ml (< 60)	• 21-hydroxylase 57.6 (<10) • adrenal cortex 1:32 (<1:10)

No antibodies found in the donor

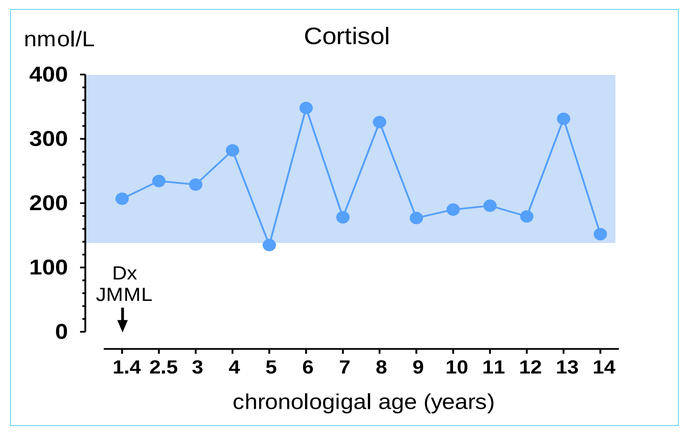

Fig. 4: Serum cortisol levels (nmol/L) during follow-up. The levels were measured at different times during the day.

References

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Adrenal

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