

Angioimmunoblastic T-cell lymphoma (AITCL) Service

Background

Lymphoma is a cancer of the lymphatic system affecting cells of the lymph nodes. Variants driving this cancer cause uncontrolled proliferation of neoplastic B and T lymphocytes which drive tumour growth. Lymphomas are classified into various subtypes based on factors such as morphology, immunophenotype, genetic markers and clinical features to aid diagnosis and treatment pathways.

Angioimmunoblastic T-cell lymphoma (AITCL) is a common subtype of peripheral T-cell lymphoma (PTCL). The incidence of AITCL increases with age, with the median age at onset reported to be 59-65 years (Fukumoto et al. 2017).

Evidence in the literature shows that variants in four specific genes (RHOA, TET2, IDH2 and DNMT3A) that are frequently seen in combination, are present at relatively high frequencies and are clinically significant for T-cell lymphomas (Lewis et al. 2020; Willemsen et al. 2018).

Test information

A next generation sequencing (NGS) panel is available for the coding regions of the following genes:

- DNMT3A – whole gene
- TET2 – whole gene
- IDH2 – Hotspot analysis of codon 172
- RHOA – Hotspots p.G17V, p.T19I and p.K18N

AWMGS will offer non-interpretative reports of results. Polymorphisms will not be reported. Results must be interpreted in combination with clinical, immunological and immunophenotypic data.

Please note that this test is not accredited by UKAS.

Referral Criteria

This service is available for diagnosis of angioimmunoblastic T-cell lymphoma for patients with confirmed T Cell Non-Hodgkin Lymphoma only. Requests should be made on the AITCL service request form available on the AWMGS website:

<https://medicalgenomicswales.co.uk/index.php/download-services>

Contact details

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Accredited to ISO 15189:2012 (8988)

Sample requirements

FFPE scrolls (see the AITCL request form for further details)

Please label samples with three identifiers and date of collection

All samples must be accompanied by a completed request form

Consent for testing and sample storage is assumed when the request is received – it is the responsibility of the referring clinician to ensure that appropriate consent has been obtained.

TAT (Calendar days) 14CD

References

Fukumoto, K., Nguyen, T. B., Chiba, C. and Sakata-Yanagimoto, M. (2017). Review of the biologic and clinical significance of genetic mutations in angioimmunoblastic T-cell lymphoma. *Wiley Cancer Science*. 109:490–496. DOI: 10.1111/cas.13393

Lewis, N. E., Petrova-Drus, K., Huet, S., Epstein-Peterson, Z. D., Gao, Q., Sigler, A. E., Baik, J., Ozkaya, N., Moskowitz, A. J., Kumar, A., Horwitz, S. M., Zhang, Y., Arcila, M. E., Levine, R. L., Roshal, M., Dogan, A and Xiao, W. Clonal hematopoiesis in angioimmunoblastic T-cell lymphoma with divergent evolution to myeloid neoplasms. (2020) *Blood Advances*. 4(10):2261-2271. doi: 10.1182/bloodadvances.2020001636.

Willemsen, M., Abdul Hamid, M., Winkens, B. and Zur Hausen A. Mutational heterogeneity of angioimmunoblastic T-cell lymphoma indicates distinct lymphomagenic pathways. (2018) *Blood Cancer Journal*. 8(1):6. DOI: 10.1038/s41408-017-0047-2.