DIAGNOSIS

In view of the histomorphologic and immunohistochemical evaluation, a diagnosis of Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) was given.

DISCUSSION

The categorization of MEITL as a distinct entity in the recent WHO classification is an acknowledgment to various works, substantiating its unique pathological and epidemiological features. Given that this neoplasm is rare, it has been known by different names in the past. For example, enteropathy associated T cell lymphoma (EATL) type II, CD56+ intestinal lymphoma and primary intestinal NK-like cytotoxic T-cell lymphoma are some of the terms used. Now, it is formally designated as monomorphic epitheliotropic intestinal TCL, giving clarity for further studies and recommendations.

The small intestine is the single most commonly involved site of MEITL. It shows no clear association with celiac disease, and appears to have increased incidence in Asians, and Hispanic populations.[4] Gastrointestinal symptoms in the cases of MEITL are often not apparent until fairly late in the disease, with perforation being a common initial presentation.[2] When diarrhoea is present, the history is measured in months rather than years; reflecting the effects of the lymphoma rather than pre-existing enteropathy. Endoscopic findings may include nodules, masses, strictures, and ulcers.[3]

On histopathological examination, the tumour features monomorphic T-lymphocytes and lacks the prominent inflammatory infiltrate that accompanies the classical EATL. The neoplastic cells have medium-sized, round nuclei with a rim of pale cytoplasm. They usually infiltrate vertically beyond muscularis propria but often there is also a radial growth pattern showing intra-epithelial spread and this epitheliotropism is a prominent feature in MEITL.[3] It is usually positive for CD3, CD8, CD56, and megakaryocyte-associated tyrosine kinase (MATK).[1] Many cases of MEITL are derived from γδ T cells, but exceptions exist; some cases are T-cell receptor (TCR) silent and some cases express TCR αβ.[2] Mutations in STAT5B JAK3 and G protein coupled receptor are associated with cases of MEITL.[5] MYC amplification (8q24) is also commanly found in these cases.[2]