E-PS-21-023
Granulomatous lymphocytic interstitial lung disease (GLILD) in CTLA-4 deficiency: case report
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Background & objectives: CTLA-4 deficiency was first described in 2014, currently categorized as immune dysregulation, with autoimmunity, immunodeficiency and lymphoid destruction (IDAIL). One of the main clinical manifestations is granulomatous interstitial lymphocytic lung disease (GLILD).
Methods: We report a case of 20 year old woman with history of multiple lymphadenopathies, ITP, hemophagocytic syndrome, and recurrent infections. A CT scan was performed due to fever and poor condition, showing bilateral pulmonary nodules, suggestive of lymphomatoid granulomatosis, deciding to perform lung biopsies.
Results: Histologic examination revealed the presence of lymphoid infiltrates with peribronchiolar distribution, forming nodular lymphoid hyperplasic aggregates (in pseudotumor pattern lesions). Abundant histiocytes and multinucleated giant cells without granulomas nor Masson bodies were found. Immunohistochemical stains confirm mixed lymphoid cellularity B (CD20+) and T (CD3), predominantly CD4+ T-cells. EBER was negative. Microbiological elements were not identified with histochemical techniques and the study of IGH and TCR-gene rearrangements was polyclonal. Given the clinical suspicion of immunodeficiency, Sanger sequencing was completed, detecting a heterozygous mutation in the germ line of the CTLA4 gene. Final diagnosis was granulomatous lymphocytic interstitial lung disease (GLILD).
Conclusion: GLILD is characterized by the presence of noncaseating granulomas and recurrent lymphoid. However, there is no consensus regarding diagnostic and therapeutic criteria, with a wide spectrum of histological patterns with or without the presence of granuloma being described in the literature, including diffuse and nodular lymphoid hyperplasia, non-follicular lymphoid aggregates, follicular bronchiolitis and organizing pneumonia, like the ones we can observe in our case. As it is a non-specific entity with low reproducibility, the term of GLILD is not recommended.

E-PS-21-024
Correlation of programmed Death Ligand-1 (PD-L1) expression with clinicopathological features in lung carcinoma in a Macedonian population
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Background & objectives: : Programmed death ligand 1(PD-L1) expression is a predictive biomarker of the success of immunotherapy for lung cancer(LC) patients, yet its prognostic significance remains unclear. This study aims to determine the relationship between PD-L1 expression and clinicopathological features in LCpatients.
Methods: The expression of PD-L1 protein in 63 surgically resected LC was evaluated by immunohistochemistry using clone 22C3 (Agilent, DAKO). The PD-L1 expression was determined by the Tumour Proportion Score (TPS) and classified as negative (TPS<1%), low-expression (TPS=1-49%) and high-expression (TPS≥50%). The statistical significance of the correlation between the clinicopathological features and PD-L1 expression was determined by chi-square test.
Results: Our study group comprised 52 male and 11 female patients, with a median age of 64 (range,33-77). 33(52.4%) of the patients exhibited PD-L1 immuno-positivity, with 23(36%) of them having a low-expression and 10(16.6%) having a high-expression of PD-L1. PD-L1 immunopositivity was significantly higher in squamous cell carcinomas (18/25;72%) compared to adenocarcinomas (10/25;40%)(p=0.023). PD-L1 expression was associated with the smoking status (p=0.0086) for the patients smoking more than 10 cigarettes per day, as well as with a higher level (>30%) of stromal tumour infiltrate lymphocytes (TILs) (p=0.049). No correlation was found between PD-L1 expression and other parameters such as patients' age, gender, stage, tumour status, grade, lymph nodal status and lymphovascular invasion.
Conclusion: This is the first local study to describe PD-L1 expression and its association with clinicopathological features in LC patients. Our preliminary results indicate that the PD-L1 protein expression in LC is associated with some clinicopathological characteristics, such as smoking status, histological type (higher expression in squamous cell carcinomas) and higher level of TILs. Further research should be performed to clarify the clinical relevance and prognostic significance of PD-L1 in LC patients.

E-PS-22 | E-Posters Soft Tissue and Bone Pathology

E-PS-22-001
Infantile fibrosarcoma with EGFR rearrangement
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Background & objectives: Infantile fibrosarcoma is a malignant fibroblastic tumour of infants. It is a rapidly growing, locally aggressive tumour characterized by t(12;15)(p13;q25), resulting ETV6-NTRK3 fusion gene. Rare fusion partners of NTRK3 or NTRK1 fusions were also reported in infantile fibrosarcoma.
Methods: RNA was isolated from 50-µm formalin fixed paraffin embedded tissues for Archer™ FusionPlex Sarcoma v2 targeted sequencing assay (ArcherDX, USA). This assay targeted fusions / mutations in 63 genes. A prepared library was sequenced using Illumina (NextSeq 500 Illumina Inc.) Next-Generation Sequencer (NGS). Produced libraries were analysed for presence of relevant fusion with the Archer analysis software version 6.2.7.
Results: A three-month-old female patient presented with a mass in the right axillary region. CT examination revealed a soft tissue mass in axillary lodge, with invasion to ribs. Gross examination revealed a firm, tumour with the largest diameter of 5,4 cm. Microscopic examination revealed a hypercellular tumour composed of fascicles of relatively uniform spindle cells with mild atypia with frequent mitotic figures. Focal herringbone pattern was present. NGS showed no mutations in NTRK gene while rearrangement was observed in EGFR gene (fusion between exon 18 and exon 25). The case was reported as infantile sarcoma with EGFR mutation. This mutation has been reported only in four patients of infantile fibrosarcoma previously.
Conclusion: Infantile fibrosarcoma is a tumour classically known to be characterized by ETV6-NTRK3 gene fusion, may have other genetic mutations including EGFR gene rearrangement. Prognostic significance of this new mutation is yet unknown.

E-PS-22-002
Extraskeletal myxoid chondrosarcoma with novel NR4A3-PRRC1 fusion