Mesenchymal tumor

Mesenchymal tissue neoplasms are soft tissue tumors, also known as connective tissue tumors, which are relatively frequent in domestic animals and have a high incidence in some species. These tumors may be located in all organs, with a higher or lower incidence in some tissues.

Phosphaturic mesenchymal tumor.

Mesenchymal tumors of the central nervous system (CNS) include numerous entities, with different pathological features and biological behavior. Mesenchymal non-meningothelial tumors are rare and comprise neoplasms that are exclusive to the CNS or show peculiar features when occurring in the CNS compared with other sites. Within this group there are three new entities, classified on the basis of specific molecular alterations and included in the 5th edition of the WHO Classification of CNS Tumors: primary intracranial sarcoma; DICER1-mutant; CIC-rearranged sarcoma; intracranial mesenchymal tumor, FET::CREB fusion-positive. These tumors often show variable morphology, making diagnosis very challenging, although the implementation of molecular techniques has led to better characterization and more precise identification of these entities. However, many molecular alterations have yet to be discovered and some recently reported CNS tumors are currently missing an appropriate classification.

Case reports

A 43-year-old man who presented with an intracranial mesenchymal tumor. Histopathological examination showed a wide spectrum of peculiar morphological features and a non-specific immunohistochemical profile. Whole transcriptome sequencing revealed the presence of a novel genetic rearrangement involving COX14 and PTEN genes, which has never been reported before in any other neoplasm. The tumor did not cluster in any defined methylation class of the brain tumor classifier, but resulted in a calibrated score of 0.89 for the methylation class “Sarcoma, MPNST-like”, when analyzed by the sarcoma classifier. The study is the first to report about this tumor with unique pathological and molecular features, characterized by a novel rearrangement between COX14 and PTEN genes. Other studies are necessary in order to define it as a new entity or as a novel rearrangement involving recently described and incompletely characterized CNS mesenchymal tumors [1].

1)
d'Amati A, Gianno F, Scuccimarri L, Lastilla M, Messina R, Signorelli F, Zimatore DS, Barresi S, Miele E, Alaggio R, Rossi S, Maiorano E, Ingravallo G, Giangaspero F, Antonelli M. Intracranial mesenchymal tumor with (novel) COX14::PTEN rearrangement. Acta Neuropathol Commun. 2023 Jun 13;11(1):95. doi: 10.1186/s40478-023-01596-9. PMID: 37312212.