INCIDENCE AND CSF BIOMARKERS PATTERN IN FRONTOTEMPORAL LOBAR DEGENERATION (FTLD) IN THE SALENTO AREA PUGLIA

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Introduction

Frontotemporal lobar degeneration (FTLD) is a focal neurodegenerative disease with progressive atrophy of the frontal and temporal lobes. FTLD includes a wide spectrum of heterogeneous clinical conditions.
The A/N biomarker classification scheme has recently been proposed, aiming to be easily applicable on an individual level. The "A" class corresponds with an amyloid beta (Aβ) and "N" with a neurodegeneration biomarker.

Objectives

The goals of the present study were:

- to assess the incidence of Frontotemporal Lobar Degeneration (FTLD) spectrum disorders in a general population of the province of Lecce;
- to define the frequencies of different FTLD phenotypes;
- to evaluate the prevalence of the neurodegeneration as assessed by Cerebrospinal Fluid (CSF) Total Tau (t-Tau) biomarker

Materials & Methods

All new cases diagnosed with FTLD spectrum disorders (incident cases), resident in the Salento Area, Puglia, inhabitants 795.134, ISTAT DATA 2019) from January 1, 2019, to December 31, 2019, were considered. The CSF biomarkers (β-Amyloid, t-Tau, p-Tau) analysis was performed in incident cases. These biomarkers were used to assess the A+/A− and the N+/N−. Based on the results of the CSF biomarker measurements, each patient was assigned to one of the four patterns of A and N.

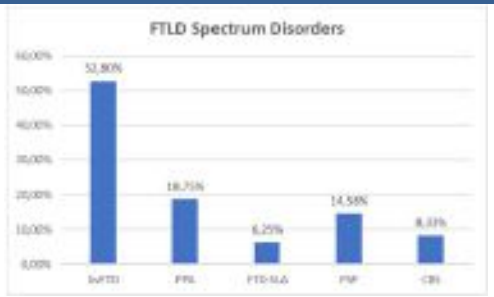

Figure 1. The figure shows the percentages of FTLD spectrum disorders.

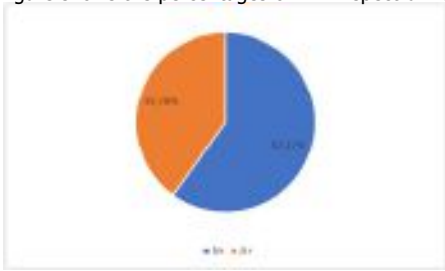

Figure 2. The figure shows the percentages of patients presented neurodegeneration (blue) and amyloidosis (orange).

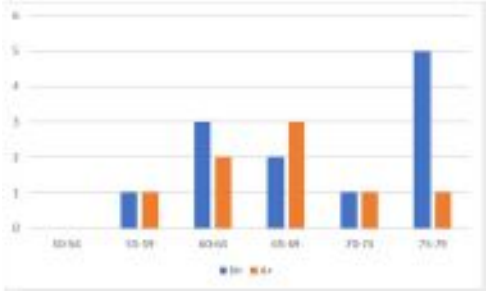

Figure 3. Frequency of neurodegeneration and amyloidosis in 5 years range of patients age.

Results & Discussion

48 patients (mean age: 70.54 years; range: 50-85) were diagnosed.
Incidence rate for FTLD was 6.03 per 100.000 person-years (py).
The behavioural variant of frontotemporal dementia (bv-FTD) was the most common phenotype (52.08%), followed by Primary Progressive Aphasia (PPA) (18.75%), Progressive Supranuclear Palsy (PSP) (14.58%), Corticobasal Syndrome (CBS) (8.33%), Frontotemporal Dementia and Amyotrophic Lateral Sclerosis (FTD-SLA) (6.25%) (Fig.1).
Of all patients, 23 (47.91%) underwent lumbar puncture; of these, 12 subjects (52.17%) presented neurodegeneration. The amyloidosis pattern was observed in 8 (34.78%) patients (Fig.2)
Stratifying by age the highest frequency of neurodegeneration was in a range of 75-79 years (21.74%) (Fig.3).
Considering the pattern in the different clinical phenotype, the neurodegeneration was present predominantly in PPA (26.08%) and bvFTD (17.39%).

Conclusions

The incident rate found in the Salento region in the year 2019 was higher than previously reported in a study conducted in 2017, on the general population of the same area [1].

The frequency of tau-related neurodegeneration was present in 52.17% of the patients evaluated. The presence of amyloidosis was detected, and this could be explained by neuropathological comorbidity due to aging or by diagnostic misclassification.

[1] Logroscino G. et al. Incidence of frontotemporal lobar degeneration in Italy. The Salento-Brescia Registry study. Neurology 2019;92:e2355-e2363.