Background: Early onset dementia (EOD) is defined as dementia with symptoms onset before 65 years. Little is known about the etiological role of environmental and occupational risk factors. We aimed at assessing the role of these factors in disease etiology. Methods: Using a case-control design, we recruited all EOD cases resident in Modena province from October, 2016 to October, 2019, as well as a referent population drawn from patients’ care-givers. We investigated residential history of study participants, and their occupational and environmental exposures to pesticides, solvents and metals through a self-administered questionnaire. We computed the odds ratios (ORs) of disease risk, and the corresponding 95% confidence intervals (CIs), according to exposure to the investigated risk factors, using an unconditional logistic regression model adjusted for sex, age, and education. Results: Fifty-eight EOD cases and fifty-four controls agreed to participate. Among occupational factors, disease risk was associated with exposure to aluminum (OR 2.6, 95% CI 0.4-15.7), pesticides (OR 2.3, 95% CI 0.7-7.8) particularly from agricultural occupational exposure (OR 3.1, 95% CI 0.7-13.3) and dyes, paints or thinners (OR 1.7, 95% CI 0.6-5.0). Among lifestyles factors, smoking (OR 1.3, 95% CI 0.6-2.9) and playing football (OR 2.2, 95% CI 0.5-9.3) or cycling (OR 2.3, 95% CI 0.4-13.4) were associated with higher EOD risk, although overall sports practice appeared to be protective factor (OR 0.4, 95% CI 0.2-0.9). Risk was also positively associated with history of head trauma (OR 1.2, 95% CI 0.3-4.1) and particularly upper arm trauma (OR 2.2, 95% CI 0.7-7.5), but not overall trauma. No association emerged for exposure to electromagnetic fields. Conclusions: Despite the study limitations, our results appear to support a role of environmental risk factors in EOD etiology, particularly of some chemical exposures and professional sports, while overall sports practice may have a beneficial effect.