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Eurofibromatosis Type-1 (NF1) is the most common autosomal dominant neurocutaneous syndrome and affecting approximately 1 in 2700 newborns.1 NF1 has high variability of expression, for that reason different clinical manifestations are seen on patients with the same NF1 gene mutation. Unidentified bright objects (UBOs) are the most common neuroimaging feature of NF1 and usually expected to disappear in adulthood. In our case, we wanted to draw attention to the fact that some of the UBOs can continue in adulthood without any clinic signs.

Neurofibromatosis Type-1; unidentified bright objects; advancing age; cognitive dysfunction

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Correspondence: Gizem GÜRSOY
Şemdinli State Hospital, Neurology, Hakkari, TURKEY
dr_gzm@hotmail.com

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CASE REPORT

Buse Rahime HASIRCI BAYIR1, Gizem GÜRSOY2, Mehmet GENCER2, Şirin YAŞAR PEKCAN3, Hülya TİRELİ3

1 Haydarpaşa Numune Training and Research Hospital, Neurology Department
2 Şemdinli State Hospital, Neurology Department
3 Haydarpaşa Numune Training and Research Hospital, Dermatology Department

Neurofibromatosis Type-1 with Unidentified Bright Objects in Advancing Age

Neurofibromatosis type 1 (NF1) is the most common autosomal dominant neurocutaneous syndrome. Unidentified bright objects (UBOs) are the most common neuroimaging feature of NF1 and usually expected to disappear in adulthood. In our case, we wanted to draw attention to the fact that some of the UBOs can continue in adulthood without any clinic signs.

Keywords: Neurofibromatosis type 1; unidentified bright objects; advancing age; cognitive dysfunction

52-year-old right handed man referred for the complaint of dizziness to Haydarpaşa Numune Training and Research Hospital Neurology Service. He did not have NF1 diagnosis before. However, he represented clinical features compatible with NF1 such as axillary and inguinal freckling, café au lait spots, Lisch nodules, neurofibromas. In family history, his mother, who had died, had neurofibromas and other clinical findings not known. As a result of evaluations, NF1 was detected in his two sisters and daughter who were not diagnosed before. In neurological examination, he had a complaint of dizziness which increased with movement, but the complaint was existed at rest. Cognitive functions, cerebellar tests and the rest of examination was normal, he had no ataxia or gait abnormalities.
Magnetic resonance imaging (MRI) demonstrated hyperintense foci, termed UBOs or focal areas of signal intensity (FASI) on T2 weighted Figure 1, and fluid attenuated inversion recovery (FLAIR) MRI scans (Figure 2). They were isointense on T1 Figure 3, and without contrast enhancement (Figure 4). UBOs were found in cortico-subcortical region of fronto-parietal white matter. A consent form was obtained from the participant.

**DISCUSSION**

NF1 is related with somewhat neuroimaging findings such as white and gray matter volumetric changes. UBOs are the most common neuroimaging feature in NF1 patients which frequently presented in thalamus, internal capsule, basal ganglia, brain stem, cerebellum and subcortical hemispheric white matter. These lesions indicate high signal on T2 and FLAIR sequences, isointense to hyperintense on T1, no contrast enhancement and no mass effect. UBOs are defined by vacuolar/spongiotic changes of myelin without inflammation and no marked demyelinization in the around tissue. UBOs noted benign and generally transient which happened in patients with 4-12 aged. Expecting to regress in adulthood and not associated
with focal neurological deficits, some persist and their presence, number and location correlated with cognitive dysfunction in recent studies. Barbier et al. reported that in 50% to 100% of the NF1 patients, MRI shows hyperintensities on T2-weighted sequences which are associated with learning disabilities. In the study of Cabellero et al., 31 patients with the NF1 were evaluated, 10% of patients had mild intellectual disability and UBOs were found in most of this subgroup. In our case UBOs were found in cortico-subcortical region of fronto-parietal white matter. Despite being seen in advanced age, they did not cause cognitive dysfunction. Mini Mental State Examination (MMSE) score was 30.

Optic nerve gliomas, sphenoid wing dysplasia, parenchymal gliomas, dural ectasia of the optic nerve sheath and spinal canal are the common manifestations central nervous system manifestations which were not seen in our patient.

Our case highlights the importance of presence, number and location of UBOs in advancing age without cognitive dysfunction. Long term prospective studies can express the reasons of regression of UBOs as age progresses and their relation with cognitive functions.

**Source of Finance**

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

**Conflict of Interest**

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

**Authorship Contributions**

**Idea/Concept:** Buse Rahime Hasırcı Bayır; **Design:** Gizem Gürsoy; **Control/Superision:** Mehmet Gencer; **Data Collection and/or Processing:** Şirin Yaşar Pekcan; **Analysis and/or Interpretation:** Hülya Tireli; **Literature Review:** Buse Rahime Hasırcı Bayır; **Writing the Article:** Buse Rahime Hasırcı Bayır; **Critical Review:** Hülya Tireli; **References and Fundings:** Gizem Gürsoy; **Materials:** Gizem Gürsoy.

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