Neuronal Lipofuscinosis in a Hairy Goat*

Hamdi AVCİ¹, Sümbül Serap BİRİNÇİOĞLU², Erkmen Tuğrul EŞİKİMEN¹, Ahmet AYDOĞAN³

¹Aydın Adnan Menderes University, Faculty of Veterinary Medicine, Department of Pathology, Aydın-TURKEY
²Special A Pathology Laboratory, İzmir-TURKEY
³Çukurova University, Faculty of Ceyhan Veterinary Medicine, Department of Pathology, Adana-TURKEY

Corresponding author: Ahmet AYDOĞAN; E-mail: aaydogan@cu.edu.tr; ORCID: 0000-0003-2504-8150


Summary: In the present case, neuronal lipofuscinosis was determined in a three-year-old female hair goat. The systemic necropsy was performed. Tissue sections were stained with hematoxylin and eosin (HE), periodic acid Schiff stain (PAS), Schmorl’s and long Ziehl-Neelsen (ZN) methods for lipofuscin; Turnbull’s blue method for hemosiderin and melanin removal method II for melanin. Unstained deparaffinized sections were examined microscopically for fluorescence in transmitted ultraviolet light. In addition, samples of pons and medulla oblongata were collected and processed for transmission electron microscopy investigations. Macroscopically, the meninges were opaque and the cerebral hemispheres had firm consistency. Microscopically, pigment granules in varying intensity were observed in the cytoplasm of the neurons of the pons and medulla oblongata. Red or yellowish-brown granules were usually seen in perinuclear localization, and filled all cytoplasm of some neurons. Moreover, meningeal and perineuronal edema, perivascular hemorrhage and neuronal degeneration were prominent histopathologic findings. The granules were positively stained with PAS, Schmorl’s and long ZN methods, but negatively stained with Turnbull’s blue method and melanin removal method II. Pigments found in all affected neurons were brightly autofluorescent. Electron microscopy also confirmed that the pigment granules were lipofuscin.

Key words: Neuronal lipofuscinosis, pathology, electron microscopy, goat

Bir Kıl Keçisinde Nöronal Lipofusinozis


Anahat kelimeler: Nöronal lipofusinozis, patoloji, elektron mikroskop, keçi

Introduction

Lipofuscin is primarily composed of cross-linked protein residues and is known as age related pigment due to lipid destruction in cell membranes with accumulation as a result of long-term and continuous autooxidation of unsaturated lipid precursors. This pigment shows intralysosomal, perinuclear location and accumulates in cells and organs with high metabolic activity such as neurons, all muscle types, liver and heart (Glees and Hasan, 1976; Cheville, 1983; Huxtable et al., 1987). It is not destroyed by the cell's proteolytic system, nor can it be removed from the cell by exocytosis. One of the most important features of lipofuscin is autofluorescence character in unstained sections by visible microscope (Cheville, 1983). In light microscopic examinations, lipofuscin pigment shows irregular granular structures in colors ranging from golden yellow to dark brown in hematoxylin and eosin (HE) stain (Carson and Hladik, 2009). In ultrastructural studies, it is seen as granules filled with vacuoles and lipid globules (Glees and Hasan, 1976; Cheville, 1983; Culling et al., 1985).
In this case report, neuronal lipofuscinosis was firstly identified with pathological and electron microscopic findings in a hair goat (*Capra hircus*).

**Case**

A three-year-old, female, hair goat was presented to pathology laboratory for necropsy. The systemic necropsy of animal was performed. Tissue samples taken after necropsy were fixed in 10% buffered formalin, processed routinely and embedded in paraffin wax. Sections were cut at 5-6 µm thickness and stained with hematoxylin and eosin (HE). The selected sections were stained by the periodic acid schiff stain (PAS), Schmorl’s, Oil Red O, Sudan Black B and long Ziehl-Neelsen (ZN) methods for lipofuscin; Turnbull’s blue method for hemosiderin and melanin removal method II for melanin (Luna, 1968; Culling et al., 1985; Carson and Hladik, 2009). Unstained depafraffinized sections were examined microscopically for fluorescence in transmitted ultraviolet light (Olympus U-LH100-3). Samples of pons and medulla oblongata taken in 1 mm also collected and processed for transmission electron microscopy investigations. These tissue samples were fixed for 48 hours in a mixture of 5% gluteraldehyde and paraformaldehyde buffered with cacodil. Then, it was washed with 1% M cacodilae buffer (pH 7.4) and post-fixed in 2% osmium tetroxide. After being dehydrated in graded alcohols and passed through propylene oxide, it was blocked with Epon 812. Thin sections were stained with toluidine blue, and the areas selected from these sections were examined with Carl-Zeiss Em 9 S electron microscope after staining with uranyl acetate and lead citrate (Culling et al., 1985).

In the anamnesis, it was reported that the animal had difficulty in walking, and sometimes it could not displace. In addition, it was stated that tremors and convulsions, which started mildly, continued increasingly after a while. Macroscopically, the goat was in poor condition. The meninges were opaque and the cerebral hemispheres had firm consistency. No macroscopic findings were found in the visceral organs. How-

Discussion and Conclusion

Lipofuscin pigment accumulates in different post mitotic cells like neurons and causes some neurologic disorders due to cellular dysfunction and degeneration (Baghban et al., 2013). Although neuronal lipofuscinosis has been identified in many animal species such as horses, cattle, cats and sheep (Huxtable et al., 1987; Jolly et al., 2002; Birinciğil et al., 2005; Baghban et al., 2013), no literature information has been available regarding the occurrence in hair goats. Here, we report, to the best of our knowledge, the first case of neuronal lipofuscinosis in a hair goat. Congenital enzyme deficiencies, toxicities, severe nutritional disorders, vitamin E deficiency, starvation due to cancer or radiation therapy and aging can be considered as the causes of accumulation of lipofuscin pigment (Huxtable et al., 1987; Birinciğil et al., 2005; Wohsein et al., 2013). In this presented case, it was thought that it may be related to both age of the animal and the long-term nutritional disorder in.
relation to the cause of the lipofuscin pigment defined only in the central nervous system. The anamnesis and gelatinous changes observed in the adipose tissue of other organs were supported this view.

Neuronal seroid lipofuscinosis should be considered in the differential diagnosis of neuronal lipofuscinosis. It is reported that histochemical stainings and electron microscopic examinations are the most common methods for distinguishing these two pigments (Culling et al., 1985; Nardocci and Cardona, 1998; Tammen et al., 2001; Jolly et al., 2002). Although lipofuscin pigment shows positive staining with PAS, Schmor's, Oil Red O, Sudan Black B, Long ZN staining methods; seroid lipofuscin pigment shows negative staining with Schmor's staining method (Culling et al., 1985; Carson and Hladik, 2009; Birincioglu et al., 2012). In this report, the pigment granules were stained positively with PAS, Oil Red O, Sudan Black B, Schmor's and long ZN staining methods. Even though electron microscopic examinations show the characteristic “fingerprint” lamellar structure in the lysosomes for seroid-lipofuscin pigment (Nardocci and Cardona, 1998), in this case, the pigment was seen in a vacuolar and lobular structure for lipofuscin pigment as reported in the literature (Glees and Hasan, 1976; Jolly et al., 1995). Moreover, negative staining was seen with Turnbull’s blue method for hemosiderin and melanin removal method II for melanin.

Lipofuscin pigment accumulates cytoplasmically in cells of many organs and tissues such as central nervous system, liver, heart, kidney and intestines. This accumulation has been reported to be a yellow-brown color by many researchers in HE staining (Culling et al., 1985; Huxtable et al., 1987; Jolly et al., 2002; Birincioglu et al., 2005; Carson and Hladik, 2009). In a case report, this pigment was stained slightly in the HE method (Baghban et al., 2009). In a case report, this pigment is stained slightly in HE staining was seen with Turnbull’s blue method for hemosiderin and melanin removal method II for melanin.

In conclusion, in this case report, neuronal lipofuscinosis was identified with pathological and electron microscopic findings in a three-year-old hair goat.

References


