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Cerebellum of a puppy

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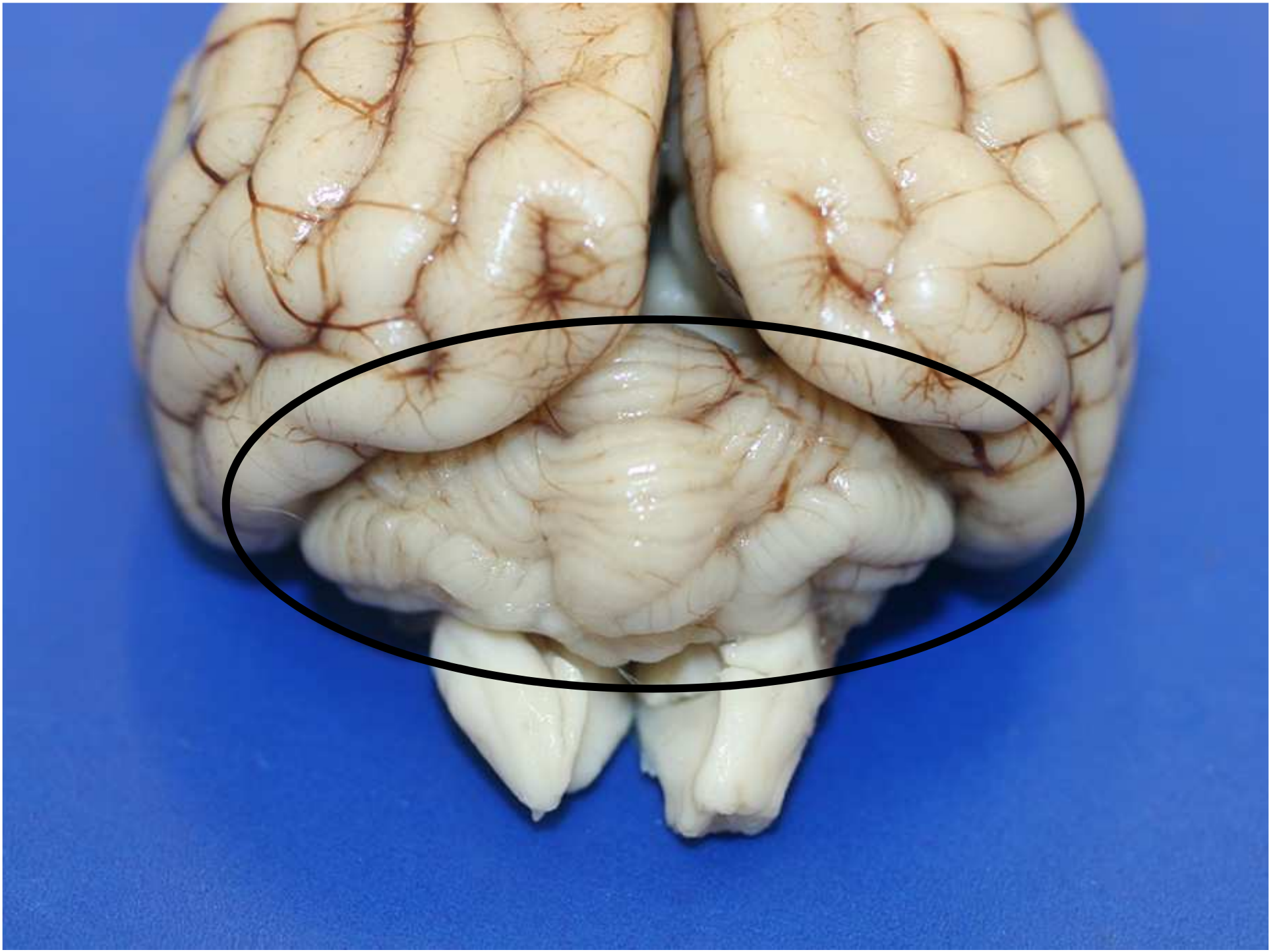
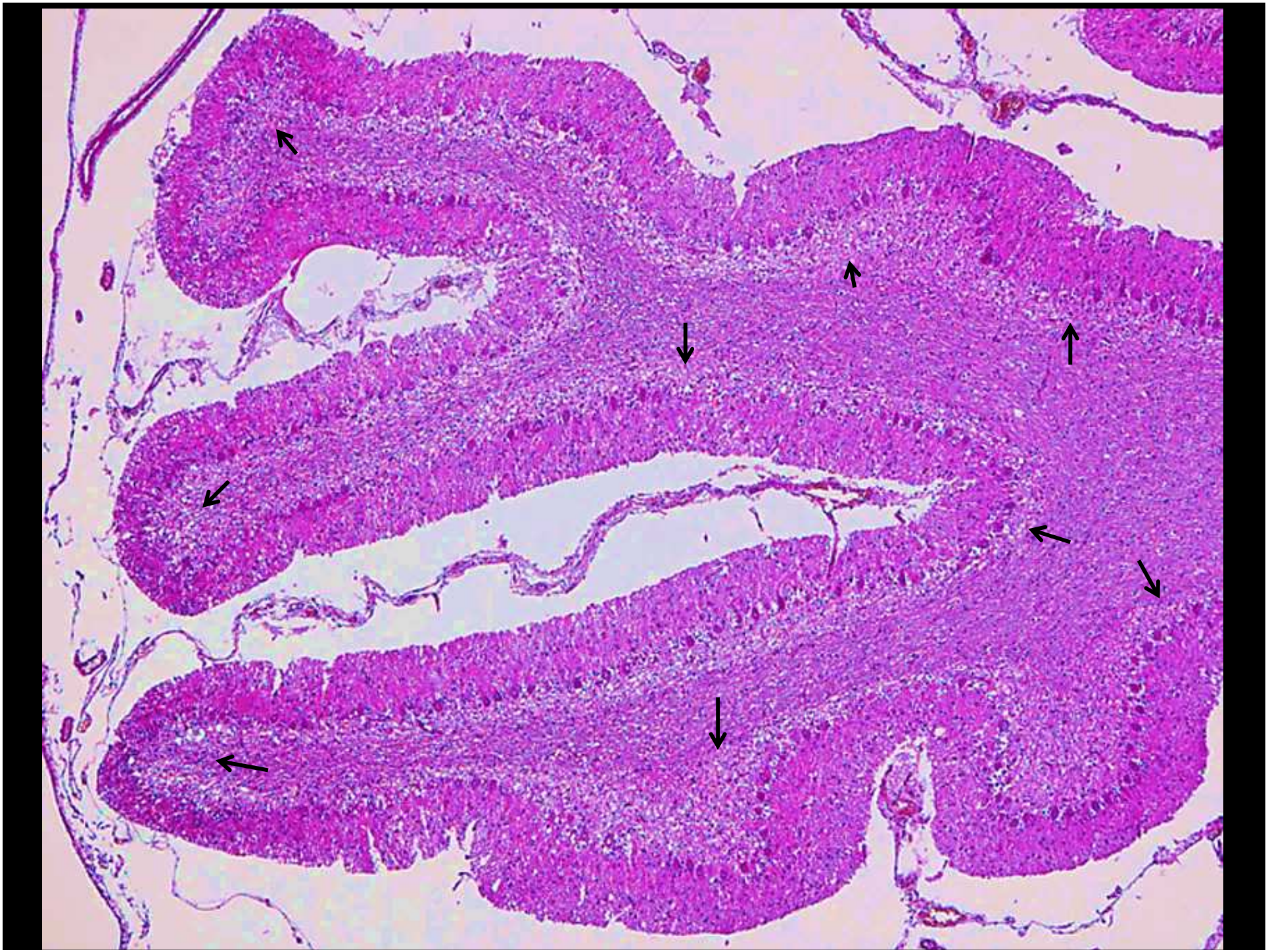
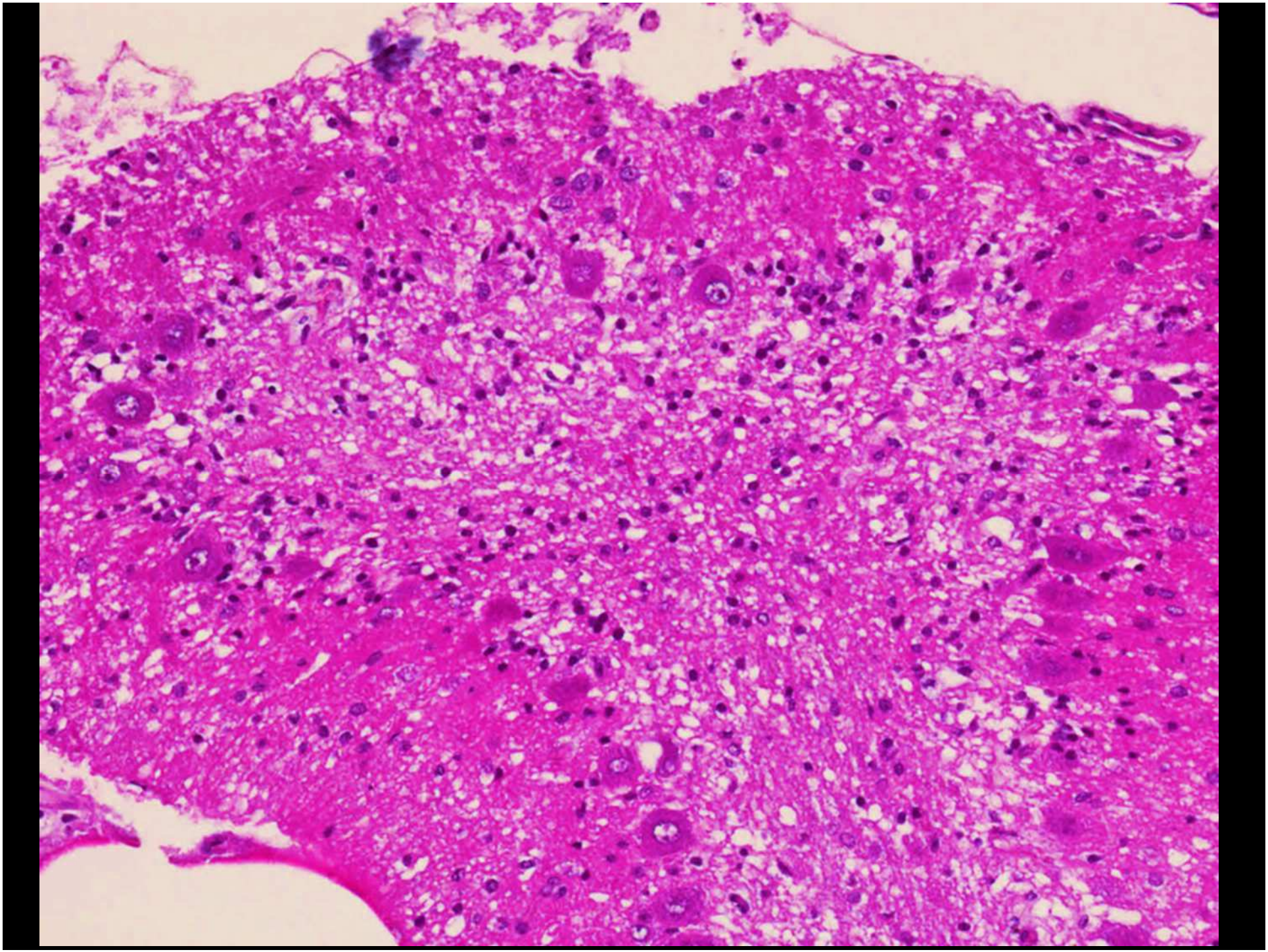
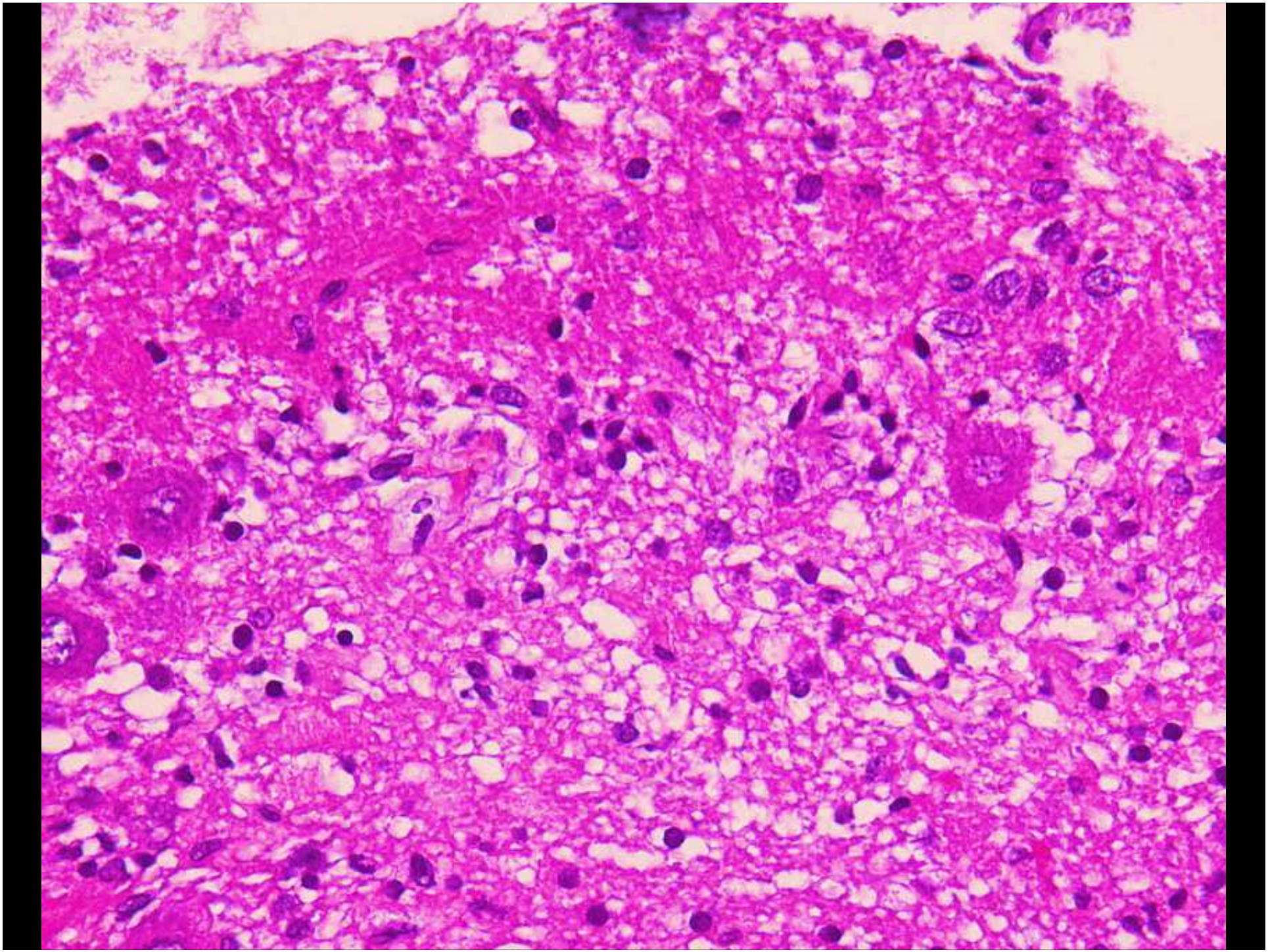
Source: infoveto.com

- **Contributor**: Vetagro Sup, France
- **Signalment:** 5 month-old, female, Coton de Tulear, dog (*Canis familiaris*).
- **History:** The dog presented with ataxia evolving since the age of 4 months originating from cerebellum on neurological examination. Magnetic Resonance Imaging revealed a decreased size of the cerebellum, without signs of inflammation.
- **Gross pathology:** Cerebellum is atrophied, slight asymmetry between its two hemispheres. The gyri were diffusely sharply delineated and shrunken.









- **Contributor's Morphologic Diagnosis:**

Cerebellum, Granular cell degeneration and loss, diffuse, severe

- **JPC Diagnosis:**

Cerebellum: Granular cell degeneration and loss, diffuse, severe, with spongiosis, and minimal multifocal Purkinje cell loss

Contributor's Comment

- **Cerebellar cortical abiotrophy** is a spontaneous, premature and progressive degeneration of neurons especially Purkinje cells with reactive gliosis without any intrinsically identifiable defect.
- Animals are born healthy and develop clinical signs after several months. A hereditary genetic defect in the glutamic acid metabolism is proposed in some dog breeds.
- **Cerebellar granuloprival degeneration** is characterized by severe depletion in the granular cell layer in this case.

- Unlike cerebellar granuloprival hypoplasia in cats caused by intrauterine **parvovirus infection**, in this case, there is no disorganization of the cerebellar cortex and no lesions in the Purkinje cell layer. Parvovirus infection in dogs does not induce cerebellar changes.
- Contrary to what has been published, in this case there is no significant inflammatory change (T cell infiltration) in the cerebellum.

Conference Comment

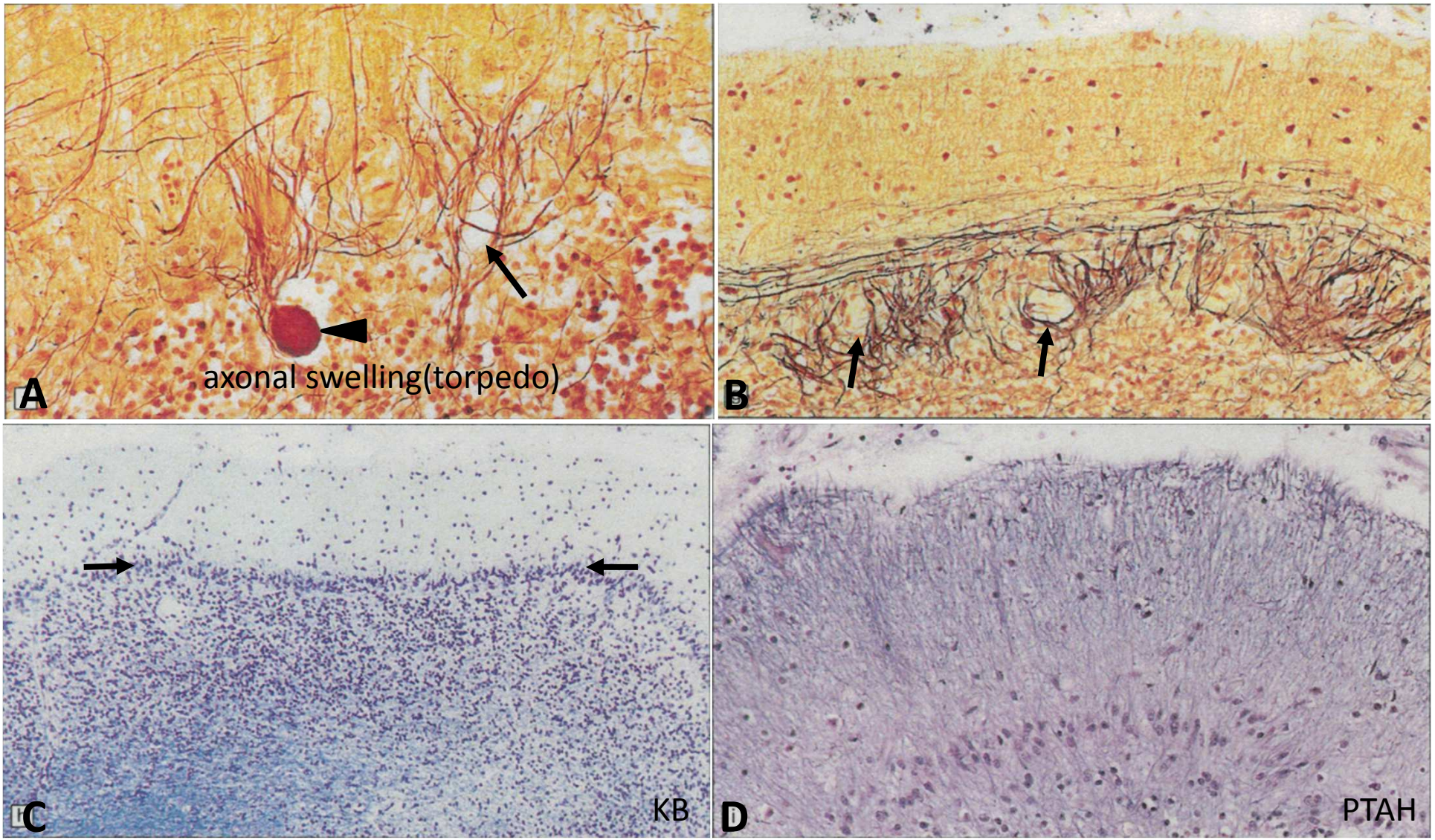
- **Cerebellar cortical abiotrophy** is hereditary defect in several breeds of dogs, Arabian horses, rabbits, an alpaca, and in goats.
- The loss of granular cell layer with only scattered loss of Purkinje cells has been reported as **cerebellar granuloprival degeneration** in a number of different canine breeds, including the Coton de Tuléar.
- Abiotrophy differs from **cerebellar hypoplasia** where the cerebellum does not completely form due to in-utero viral infections from parvoviruses or pestiviruses and certain toxicities eg. organophosphates.

- The increased number of hypertrophic astrocytes with large vesicular nuclei within the Purkinje cell layer (**Bergmann gliosis**) occurs predominantly in areas where Purkinje cells are lost, described as **empty baskets**.
- Pathogenesis of cerebellar granuloprival degeneration in this breed is unknown. An inherited disorder of granular cell development is hypothesized, but most Purkinje cells survive since their main excitatory input is from the olivary nucleus.
- Purkinje cells could be lost as result of granular cell depletion in chronically affected dogs

Empty basket and Bergmann gliosis in Cerebellar cortical degeneration.

(**A**), (**B**) 'empty basket' [Silver impregnation]

(**C**), (**D**) Bergmann astrocytosis.



Neuropathology, A reference text of CNS pathology, 3rd ed. pp.589 (2013)