

Pelger-Huët Anomaly in a Bitch Basenji

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ABSTRACT

Background: Pelger-Huët anomaly (PHA) is characterized by morphological changes in all granulocytes, being more evident in neutrophils. Granulocytic function in these animals remains unchanged. Hereditary form of PHA should be differentiated from pseudo-PHA, a condition acquired from infections and/or inflammation conditions. Recognition of PHA is important to avoid misleading leukogram interpretation, since hyposegmentation of neutrophils can be confused with left shift, making it necessary to carry out diagnostic tests and treatment for the disease that is generating the deviation. The objective of this case report was to demonstrate the importance of laboratory diagnosis in PHA.

Case: A 11-month-old bitch Basenji, was presented to perform preoperative evaluation for elective ovariosalpingohysterectomy at Veterinary Hospital (HUVET) of the Universidade Federal Fluminense (UFF). Tutor reported that animal was healthy, vaccination status was current, had deworming protocol applied and had not made use of medications recently. Animal presented normophagy, normodipsia, normuria and normochezia. Upon physical examination, animal was alert consciousness level, with adequate hydration, hyperemic oral mucosa, a less than 2 s capillary perfusion time, normal lymph nodes (submandibular, pre-scapular, inguinal and popliteal), rectal temperature of 39.2°C, heart rate of 160 beats per minute and respiratory rate of 60 movements per minute, possibly due to the animal's agitated state. Abdominal palpation showed no changes. Physical examination presented no alterations. Preoperative exams included complete blood count (CBC) and biochemistry profile (ALT, AP, glucose, creatinine, urea, total proteins and fractions). Samples were sent to Hospital's Veterinary Clinical Pathology Laboratory (LABHUVET/UFF) for analysis. CBC was performed using automatic method. Blood smears were stained with hematological stain and then a cytomorphological evaluation was performed. The first CBC revealed 23% of neutrophils with nuclear hyposegmentation and 44% of neutrophils were bands. A follow up was performed after 9 months, and a Complete Blood Count was performed again in which 12% of neutrophils showed nuclear hyposegmentation with mature chromatin pattern, 40% of neutrophils were bands, 1% of meta-myelocytes neutrophils, 1% of myelocytes neutrophils and, also, eosinophils with nuclear hyposegmentation. Animal was healthy, and had no alterations on physical examination suggesting a diagnosis of PHA.

Discussion: Recognition of PHA is important to avoid misleading leukogram interpretation, since neutrophils hyposegmentation can be confused with left shift, which is considered severe with poor prognosis, making it necessary to carry out diagnostic tests and treatment for the disease that is generating the deviation. The diagnosis of PHA was considered by the shape of the leukocytes nuclei, without evidence of inflammatory disease, during the patient follow up. Therefore, this anomaly should be considered as a differential diagnosis of left shift, thus avoiding unnecessary clinical and therapeutic procedures. Guidance in face of this hereditary hematological syndrome is important. The responsible guardian of the animal must not allow it to act as a breeder in order to interrupt possible transmission of this anomaly to offspring, because there is a fatal form when it comes to homozygotes.

Keywords: canine, dog, hereditary anomaly, WBC, nuclear hyposegmentation, neutrophils, left shift.

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INTRODUCTION

Pelger-Huët anomaly (PHA), an autosomal dominant hematological trait is characterized by neutrophil nuclear hypolobulation [19]. Granulocyte nuclei can be round, oval, in the shape of a dumbbell, in the shape of a peanut, bilobulated, or even as rods [11].

PHA has already been described in humans [3,21], rabbits [16], cats [15], dogs [5,13,15], horses [6,8] and mice [17]. In dogs, condition has been described in a variety of breeds, including Cocker Spaniel [5], Basenji [14], Border Collie, American and English Foxhounds [13], Shepherd Australian [12], Boston Terrier [11] and in mixed breed dogs [10].

Recognition of this anomaly is important to avoid misleading leukogram interpretation, since hyposegmentation of neutrophils can be confused with left shift, making it necessary to carry out diagnostic tests and treatment for the disease that is generating the deviation. Some morphological findings observed in the left shift such as cytoplasmic basophilia and toxic granulations are not found in PHA, highlighting differences between the two conditions [1,4,7,18].

We report a case of PHA in a bitch Basenji, domiciled in the State of Rio de Janeiro, Brazil, emphasizing cytomorphological examination of blood smear and physical examination to rule out inflammatory conditions.

CASE

A 11-month-old bitch Basenji was received to perform preoperative evaluation for elective ovariosalpingohysterectomy at Veterinary Hospital of Universidade Federal Fluminense. Tutor reported that animal was healthy, vaccination status was current, had deworming protocol applied and had not made use of medications recently. Animal presented normophagy, normodipsia, normuria and normochezia. Upon physical examination, animal was alert consciousness level, with adequate hydration, hyperemic oral mucosa, a less than 2 s capillary perfusion time, normal lymph nodes (submandibular, pre-scapular, inguinal and popliteal), rectal temperature of 39.2°C (reference values = 37.5° to 39.2°C), heart rate of 160 beats per min (reference values = 60 to 160 beats per min) and respiratory rate of 60 movements per min (reference = 18 to 36 movements per min), possibly due to the animal's agitated state. Abdominal palpation showed no changes.

Preoperative exams included complete blood count (CBC) and biochemistry profile (ALT, AP, glucose, creatinine, urea, total proteins and fractions). For this, 6.0 mL of blood was collected from jugular vein, which was immediately placed in an ethylenediaminetetraacetic acid (EDTA) tube and in a clot activating gel tube. Samples were sent to Hospital's Veterinary Clinical Pathology Laboratory (LABHUVET/UFF) for analysis. CBC and biochemistry profile were performed using automatic methods, MindrayBC-2800Vet^{®1} and Wiener Lab Group CM250^{®2}, respectively. Blood smears were stained with diff-quick stain³ (Panótico rápido[®]) and then a cytomorphological evaluation was performed. The first CBC was performed on 12th June of 2019 and the cytomorphological evaluation revealed nuclear hyposegmentation with mature chromatin pattern in 23% of neutrophils and 44% of band neutrophils presenting mature chromatin pattern (Figure 1). Nine months after the 1st examination, a new CBC was performed, in which 12% of neutrophils showed nuclear hyposegmentation with mature chromatin pattern, 40% of neutrophils were bands, 1% of metamyelocytes neutrophils, 1% of myelocytes neutrophils and, also, 11% of eosinophils with nuclear hyposegmentation. Considering the patient's health condition, a presumptive diagnosis of PHA was made. Regarding the biochemical results, the 1st evaluation showed no changes in values of urea, creatinin, alanine aminotransferase, alkaline phosphatase, total protein, albumin, globulin, albumin:globulin ratio and glucose. The 2nd evaluation revealed an increase in alanine aminotransferase (value of 677 U/L; reference values = 21 to 102 U/L [9]). During this 9-month period, the animal received no treatment or developed any disease.

The diagnosis of PHA was considered by the shape of the leukocytes nuclei, without evidence of inflammatory disease, during the patient follow up.

DISCUSSION

PHA is a rare hereditary disorder characterized by incomplete segmentation of the nucleus of granulocytes, mainly neutrophils, due to failure in nuclear maturation [20]. In this anomaly, neutrophils present hyposegmented or round nuclei with condensed chromatin and mature leukocyte cytoplasm [1,4,7,18], compatible with neutrophils morphology observed on blood smear evaluation in the reported case. Hyposegmentation occurs in 30 to 70% of neutrophils and

mimics the state of cellular immaturity that, initially, may suggest a left shift [1,22].

This anomaly is an autosomal dominant hereditary disease that presents 2 states of manifestation, heterozygous and homozygous. Heterozygous state is the most common observed and the animal has a normal life. The homozygous state of the anomaly is rare and generally lethal [2,11]. Despite change in granulocyte morphology, function of these cells is not compromised and carriers of this anomaly are no longer susceptible to infections and/or immunodeficiency syndromes [1,4,7,18]. Probably the case described is a presentation of the heterozygous form, since in a 9 month period no clinical alterations were observed and homozygous is lethal [2,11].

PHA occurs in several canine breeds such as Basenji [4,18] as described in this report.

In order to diagnosis this anomaly, differentiation must be made for other forms of nuclear hyposegmentation that are acquired, the so-called Pelger-Huët pseudo-anomalies, such as infectious, neoplastic diseases and drug exposures. For this, it is necessary to analyze 3 characteristics, first, the presence of persistent granulocyte nuclear hyposegmentation in hematological analysis. Second, exclude Pelger-Huët pseudo-anomalies capable of causing hyposegmentation such as infections and/or inflammation conditions. And, finally, investigate the transmission of the anomaly among family members [1]. This reported case cannot be confused with pseudo-anomalies since infections, neoplasms and use of drugs as a cause of persistent hyposegmentation were excluded

during the entire evaluation period. In addition, the clinical examination and the laboratory tests results confirmed that the dog was healthy. Unfortunately, it was not possible to access patient's relatives to prove a hereditary basis.

Leukocytes morphological evaluation by microscopic examination of blood smear is an important component of complete blood count. Even when total number of leukocytes is within reference range, identification of increased number of hyposegmented neutrophils (left shift) can mean an inflammatory response. Neutrophil hyposegmentation also occurs in PHA, so it is important a differential diagnosis.

Analysis of the 1st blood smear showed a large number of hypo-segmented leukocytes, with a pattern of mature chromatin, in the form of bands and metamyelocytes, presenting a deviation to the left, which did not correspond with the patient's clinical presentation. In addition, other cells in the granulocytic lineage such as eosinophils also presented hyposegmentation (Figure 1). Evaluation of a new blood smear from the same animal confirmed the result after 9 months of the first blood analysis, showing the same pattern of morphological changes in granulocytic leukocytes (neutrophils and eosinophils), and compatible with Pelger-Huët anomaly. Beside suppression of segmentation of granulopoiesis, the nuclei of all leucocytes, including neutrophils, lymphocytes, monocytes, eosinophils and basophils, are affected in PHA. As the patient did not present any clinical alterations; the Pelger-Huët anomaly was suspected.

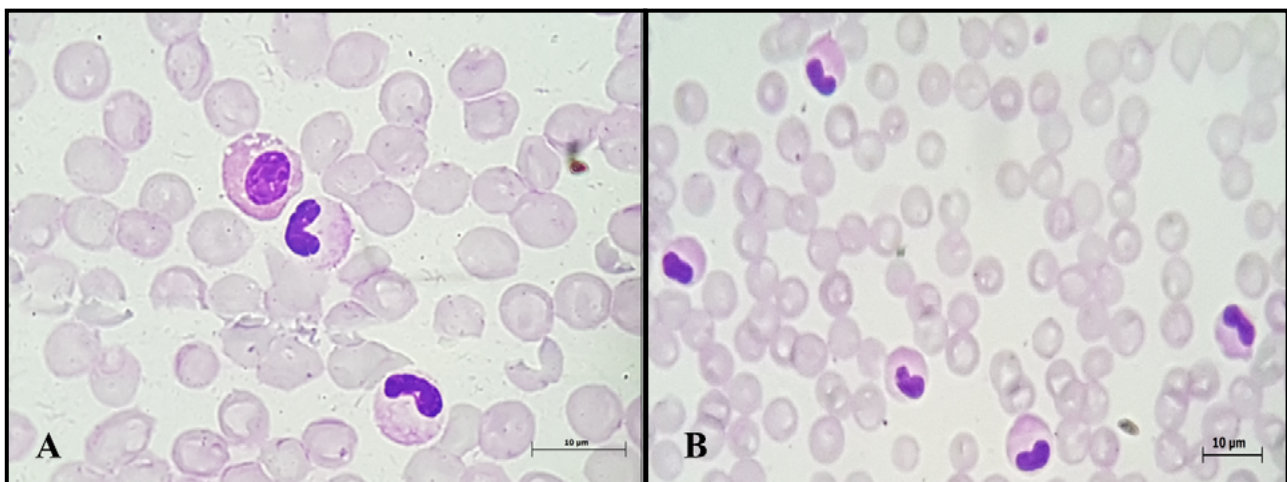


Figure 1. Blood smear from the bitch Basenji with Pelger-Huët Anomaly (different fields). Blood smear was stained with diff-quick stain (Panótico Rápido®). A- There is granulocytic hyposegmentation in two rod forms of neutrophils and in one myelocytic form of eosinophil [400x]. B- Several neutrophils presenting hyposegmentation [100x].

Guidance in the face of this hereditary hematological syndrome is crucial. Although rare and the most common manifestation is benign for the carrier, the tutor must not allow the animal to act as a breeder in order to interrupt the possible transmission of this anomaly to the offspring, because there is a fatal form when it comes to homozygous [1,7].

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Declaration of interest. The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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