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CASE REPORT
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Lateral Patellar Luxation and Ehlers Danlos Syndrome (EDS) in a Dog

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ABSTRACT

***Background*:** Ehlers Danlos Syndrome (EDS) is a rare genetic disease characterized by a deficiency in collagen synthesis, which can result in joint laxity. Patellar luxation is one of the main orthopedic conditions that affect the canine knee joint, with limited descriptions of its association with EDS in dogs. The purpose of this report is to describe the surgical management and postoperative evolution of a 1-year-old Chow Chow dog with grade II patellar luxation, tibial valgus and EDS. ***Case*:** A 1-year-old Chow Chow dog was referred to the University Veterinary Hospital due to lameness of the left pelvic for 3 months. At the orthopedic examination were verified severe lameness and lateral deviation of the left stifle joint during the ambulation of the animal. Additionally, it was verified bilateral hyperextension of the tibiotarsal joint and grade II patellar luxation of both pelvic limbs with painful hyperextension of the left stifle joint. Radiographic evaluation showed lateral displacement of the patella from both femoral trochlear groove, and a valgus deviation of the proximal left tibial shaft. In addition, it was verified cutaneous hyperextensibility and an extensibility index suggestive of EDS. The animal was submitted to trochlear block resection technique and medial imbrication, followed by corrective tibial osteotomy. Furthermore, skin biopsies of the scapular and lumbar folds were performed during the corrective tibial osteotomy. The samples were sent for histopathological examination, which revealed fragmented and unorganized collagen fibers in the dermis. Histopathological findings were compatible with EDS. The absence of lameness and correct positioning of the patella in the trochlear sulcus were verified in the post-surgical follow-up. Complete bone consolidation of the closing wedge osteotomy to correct the tibial valgus was verified at 90 days postoperatively.
***Discussion*:** The clinical signs, cutaneous extensibility index, and histopathological abnormalities in the present case were consistent with EDS. In the present study, this congenital collagen abnormality syndrome may have been a contributing factor of patellar luxation as EDS can result in hypermobility of ligaments and joints, due to metabolic and structural abnormalities of the collagen in connective tissues, and consequently may promote patellar luxation and other orthopedic abnormalities. A variant of EDS in humans has been implicated in the development of skeletal abnormalities such as short stature and bone deformities. This corroborates the possibility that EDS is correlated with valgus angulation of the proximal portion of the tibia in the present case. However, in-depth genetic studies are required to confirm this correlation. Corrective osteotomy in conjunction with block recession sulcoplasty and medial imbrication seem to have enabled patellofemoral stability and alignment of the quadriceps mechanism, ensuring that the patella remained in the trochlear sulcus, even in the presence of EDS. In addition, this syndrome does not seem to affect the surgical outcome of the treatment of patellar luxation associated with closed wedge osteotomy for tibial valgus correction. Medium-term follow-up can be considered excellent in this case report since there was a rapid resolution of lameness and adequate corrective osteotomy healing despite persistent hyperextension of the tibiotarsal joint. Ehlers Danlos Syndrome did not contraindicate the surgical treatment of patellar luxation. However, further studies are needed to assess the influence of the syndrome on long-term patellar luxation. The findings of this case report can help in the diagnosis and treatment of other animals affected by this rare syndrome and associated orthopedic diseases.

Keywords: patellar luxation, bone, collagen diseases.

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INTRODUCTION

Patellar luxation (PL) is one of the main orthopedic conditions that affects the stifle joint of dogs. PL is considered a developmental disorder because it mostly occurs after birth or early in life, although the underlying etiopathogenesis remains unclear. Indeed, this condition could be secondary to trauma and as a complication of the treatment of cranial cruciate ligament disease or fractures involving femur or tibia. Patellar luxation has been associated with proximal tibia and/or distal femur bone deviations, shallow trochlear groove and soft tissues abnormalities [5,6].

Ehlers Danlos Syndrome (EDS) is a rare genetic disease reported in humans, dogs, cats, rabbit, mink, cattle, and sheep. The disease is characterized by a deficiency in collagen fibers synthesis, which can cause cutaneous fragility, cutaneous hyperelasticity and joint laxity. This syndrome appears to be correlated with patellar luxation and disease severity in dogs [11].

Treatment of patellar luxation involves surgical correction of bone abnormalities and joint laxity. The surgical treatment can be challenging in animals with comorbidities, such as EDS [5].

The aim of the present case report is to describe the surgical technique and the postoperative follow-up of a 1-year-old Chow Chow dog with grade II patellar luxation and tibial valgus associated to EDS, submitted to closing wedge tibial osteotomy. To the best of our knowledge, there are no reports describing the surgical procedure and postoperative follow-up in a complex case of lateral patellar luxation associated with EDS in dogs.

CASE

A 1-year-old male Chow Chow dog was referred to the University Veterinary Hospital due to lameness of the left pelvic limb for 3 months. At the orthopedic examination were verified severe lameness and lateral deviation of the left stifle joint during the ambulation of the animal. Additionally, it was verified bilateral hyperextension of the tibiotarsal joint and grade II patellar luxation of both pelvic limbs with painful hyperextension of the left stifle joint.

In addition to joint laxity, it was verified cutaneous hyperextensibility. Due to this, it was measured the extensibility index and the result suggestive of Ehlers Danlos Syndrome (EDS) [11].

Radiographic evaluation showed lateral displacement of the patella from both femoral trochlear groove, and a valgus deviation of the proximal left tibial shaft (Figure 1). No more osseous deformities have been presented during exam evaluation.

The animal was submitted to trochlear block resection technique and medial imbrication [2], followed by corrective tibial osteotomy [5].

Preoperative planning to correct tibial valgus included the measurement the magnitude of the angulation from the center of rotation of angulation (CORA) on the tibial mechanical axis using mediolateral radiographic projections [8]. The magnitude of angulation of the proximal diaphyseal deformity was 20°.

For the surgical procedure, the animal was positioned in dorsal recumbency under general anesthesia and it has your left pelvic limb aseptically prepared. Then, the lateral arthrotomy of the left stifle joint was performed by a cranial access. After this procedure, it was performed trochlear block resection technique,

Figure 1. Preoperative radiographic evaluation of a 1-year-old Chow Chow with Ehlers-Danlos Syndrome (EDS). A- The craniocaudal projection shows the valgus deviation of the stifle joint and the proximal shaft of the left tibia. B- The lateral projection shows the absence of a craniocaudal deviation of the left tibia.

followed by medial imbrication using 2-0 monofilament polyamide[1]. Immediately thereafter, it was performed a cutaneous incision on the medial surface of the left tibia, in order to expose the tibial diaphysis [2]. For tibial valgus deformity, a mediolateral closing wedge was then performed using an oscillatory saw. The location of the medial closing wedge was based on the CORA and the wedge angle was determine by using a sterilized caliper as well a right triangle method [10].

After the realignment of the tibia, the wedge was fixed with double locking plate, positioned in a neutralization configuration. A 2.7 mm locking compression plate (LCP) of 6 holes[2] was positioned on the cranial surface of the tibia, while another 2.7 mm LCP of 10 holes[2] was positioned on the medial surface of the tibia. For the cranial locking plate, there were positioned 2 bicortical screws[2], respectively, in the proximal and distal segments of the osteotomy. For the medial locking plate, there were positioned 3 and 2 bicortical screws[2], respectively, in the proximal and distal segments of the osteotomy.

Surgical incision was closed in layers. In addition, skin biopsies of the scapular and lumbar folds were performed during the corrective tibial osteotomy. The samples were sent for histopathological examination (HE)[3] which revealed fragmented and unorganized collagen fibers in the dermis (Figure 2). Thus, the histopathological findings were compatible with EDS [11].

The animal received at the postoperative period tramadol hydrochloride [Tramal Retard®[4] - 4 mg/kg/PO/TID, for 7 days], cephalexin [Lexin®[5] - 30 mg/kg/PO/BID, for 7 days], meloxican [Maxicam®[6] - 0.1 mg/kg/PO/SID, for 3 days] and omeprazole [Gaviz V®[7] - 0.5 mg/kg/PO/SID, for 7 days]. During immediate postoperative radiographic evaluation were verified the alignment of the mechanical axis of the left tibia, and the adequate positioning of the metallic implants (Figure 3).

The patient presented improvement of walking at seventh postoperative day. At 15th postoperative day, the animal did not present lameness, and the patella was adequate positioned in the femoral trochlear groove. Despite this, hyperextension of the tibiotarsal joints was still verified. The tibial bone consolidation was observed at 90th postoperative day with minimal bone callus formation and with no reluxation verified (Figure 4).

DISCUSSION

In the present study, a congenital collagen abnormality known as Ehlers Danlos Syndrome (EDS) may have been a contributing cause of patellar luxation. This collagen disease can result in hypermobility of ligaments and joints, due to metabolic and structural abnormalities of the collagen in connective tissues, and consequently may promote patellar luxation and other orthopedic abnormalities [1].

The diagnosis of EDS is based on clinical signs, including cutaneous extensibility index, histopathological examination and electron microscopy [9,11]. The clinical signs, cutaneous extensibility index, and histopathological abnormalities in the present case were consistent with the disease and made possible the diagnosis of EDS. The owner of the animal declined the electron microscopy.

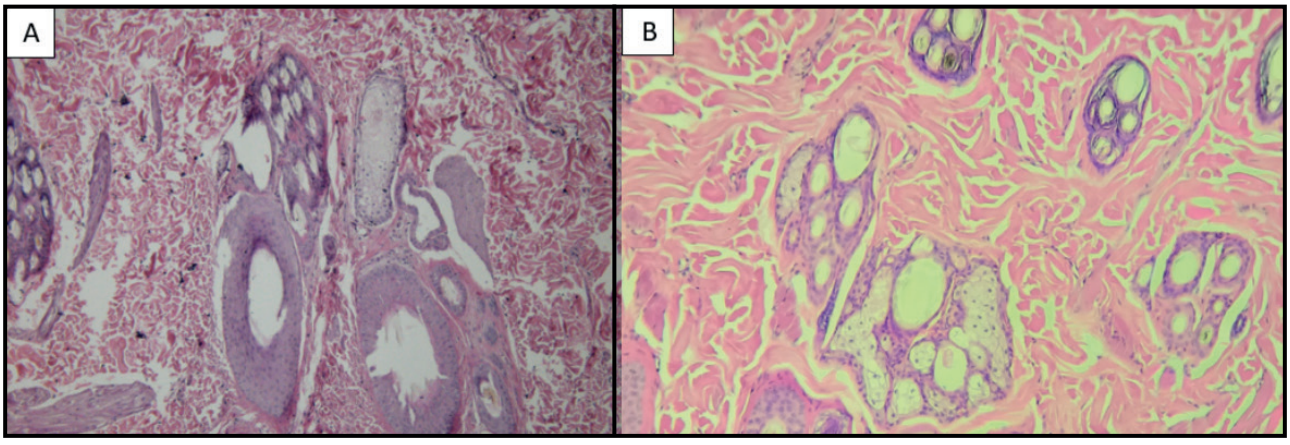

Figure 2. A- Histopathological examination of the skin of a 1-year-old Chow Chow with Ehlers-Danlos Syndrome-Like (EDS). Note elastic fibers presented but with an irregular distribution. B- Histopathological comparison with features of a skin of a healthy dog. [HE; 100x].

Figure 3. Radiographic evaluation at immediate postoperative surgical procedure of a 1-year-old Chow Chow with Ehlers-Danlos Syndrome-Like (EDS), submitted to corrective tibial osteotomy. A- The craniocaudal projection shows the alignment of the mechanical axis of the left tibia, as well as the stifle joint. Note the opening wedge at the middle shaft of the left tibia. B- The lateral projection shows the adequate positioning of the metallic implants. Note the opening wedge at the middle shaft of the left tibia.

Figure 4. Radiographic evaluation at 90 days postoperatively of a 1-year-old Chow Chow with Ehlers-Danlos Syndrome (EDS), submitted to corrective tibial osteotomy. A- The oblique projection shows the continuity of both bone cortices with minimal bone callus formation. B- The lateral projection shows bone filling of the wedge and adequate positioning of the metallic implants.

The EDS has no specific treatment. Clinical or surgical management can be considered depending on the clinical manifestations presented by the animal. Promising post-surgical results have been reported after skin resection, surgical correction of prolapsed gland of the third eyelid as well as entropion in canine patient with EDS. In addition, the surgical treatment can result in postoperative complications resulting from easily tearing skin due to collagen dysplasia leading to poor tensile strength of the skin and consequently suture dehiscence [9]. Postoperative complications, such as suture dehiscence, were not verified in the present case.

Angular deformities in limbs are mainly due to abnormalities in the growth plates due to an inherited condition or secondary to traumatic events [7]. In our case report, a cause for valgus deviation in the proximal tibia cannot be elicited. A variant of EDS in humans has been implicated in the development of skeletal abnormalities such as short stature and bone deformities. This corroborates the possibility that EDS is correlated with valgus angulation of the proximal portion of the tibia in the present case [3]. However, in-depth genetic studies are required to confirm this correlation.

Furthermore, this syndrome causes joint laxity and have been implicated with patellar luxation and disease severity in dogs [11]. Thus, the syndrome may have contributed to patellofemoral instability, allowing patellar luxation. The fact that the luxation was lateral may be related to the valgus deviation present in the tibial diaphysis, which, in turn, triggered malalignment of the quadriceps mechanism, lateral luxation of the patella and shallow trochlear groove [6].

Although genu valgum has a high incidence in dogs with lateral patellar luxation and is associated with the severity of the disease, it remains uncertain whether the genu valgum reflects an underlying bone

deformity that would influence the development of lateral patellar luxation, or whether it would be just a posture adopted by the dog to compensate for patellar instability. The retrospective nature of most studies evaluating lateral patellar luxation makes it impossible for diagnostic images of angular limb deformities to be available [5]. Thus, further studies are needed to investigate the correlation between angular limb deformities and lateral patellar luxation. The fact that the dog in our report did not present reluxation may reinforce our hypothesis that the tibial valgus was contributing to the lateral luxation of the patella. Corrective osteotomy in conjunction with block recession sulcoplasty and medial imbrication seem to have enabled patellofemoral stability and alignment of the quadriceps mechanism, ensuring that the patella remained in the trochlear sulcus, even in the presence of EDS. Although a limitation in our study was the impossibility of long-term follow-up. Additionally, other orthopedic lesions verified in the present study, such as bilateral hyperextension of the tibiotarsal joint, corroborated the diagnosis of EDS, since the collagen disease has different degrees of systemic involvement and may cause besides cutaneous hyperextensibility, vascular fragility and multiple orthopedic disorders due to joint laxity [10].

The closing wedge osteotomy requires a rigid fixation [10]. In the present case, rigid fixation was obtained by the placement of 2 locking plates. The positioning of 2 locking plates was due to the possibility of insufficient locking of the implants in the trans-operative. In a study evaluating plates constructs in single-cycle bending and torsion, the double locking fixation showed significantly greater bending stiffness, bending strength and torsional stiffness when compared to the broad single locking fixation [4]. Therefore, the medial and cranial placement of the plates was performed according to the forces to be neutralized [2]. The authors also consider that a single longer locking plate could be a viable alternative to the fixation performed in the present case.

Bilateral hyperextension of the tibiotarsal joint persisted after the corrective tibial osteotomy procedure, probably due to the EDS and consequently the joint laxity [1]. Although still impairing the ambulation, this orthopedic abnormality did not interfere with postoperative recovery and bone consolidation.

In summary, this report describes a successful correction of a lateral patellar luxation grade II with associated tibial valgus deformity and EDS in a dog. Medium-term follow-up can be considered excellent in this study since there was a rapid resolution of lameness and adequate corrective osteotomy healing despite persistent hyperextension of the tibiotarsal joint. EDS did not contraindicate the surgical treatment of patellar luxation. Thus, the findings of this case report can help in the diagnosis and treatment of other animals affected by this rare syndrome and associated orthopedic diseases.

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***Declaration of interest*.** The authors declare no conflicts of interest related to this report. The authors alone are responsible for the content and writing of paper.

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