Case report
Retrobulbar Rhabdomyosarcoma in a Hovawart

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Abstract

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This clinical case study describes a three-year-old female Hovawart dog presented due to a unilateral exophtalmos and diagnosed with a retrobulbar tumour arising from m. rectus ventralis using ultrasound and computer tomography. The tumour was cytologically classified to be of a malignant mesenchymal kind. Modification of lateral orbitotomy with canthotomy was performed and the circumscribed tumour resected in its entirety. After healing, the eye was like the healthy contralateral one. Three months later, however, there was a relapse of the same extent and the patient was euthanised on the owner´s request. Rhabdomyosarcoma was confirmed by repeated histological examinations.

Tumour, dog, orbitotomy

Retrobulbar processes belong to relatively frequent ophthalmological problems of dogs and cats. Apart from inflammatory lesions (abscesses, cellulitis), there are frequently processes of neoplastic origin such as primary or metastatic tumours (Spiess and Wallin-Hakanson 1999). Kern (1985) mentioned up to 74% of retrobulbar tumours to be of metastatic origin. Cases of retrobulbar tumours are very often malignant. Manson et al. (2001), Kern (1985), and Hendrix and Gelatt (2000) report 42%, 91%, and even up to 95% of malignant tumours, respectively. A total of 21 different retrobulbar neoplasia was described with osteosarcomas, fibrosarcomas and nasal adenocarcinomas (cf. Table) being the most frequent ones. There is one reported case of a retrobulbar rhabdomyosarcoma (Hendrix and Gelatt 2000). No age and breed predisposition of the occurrence of retrobulbar tumours was found (Attali-Soussay et al. 2001). The mean age, however, is relatively high; i.e., 8.4 and 10.7 years reported by Hendrix and Gelatt (2000) and Attali-Soussay et al. (2001), respectively. Kern (1985) mentions purebred dogs, mostly females of middle age, to be typically affected. Prognosis of cases of retrobulbar neoplasia is generally poor with low survival of patients. Attali-Soussay et al. (2001) mention that dogs survive for up to 10 months. According to Hendrix and Gelatt (2000) 86% and 19% of dogs live longer than 6 and 12 months, respectively. Radical treatment accompanied by a partial or total orbitectomy decreases the relapse of tumours to 36.7%. More than 70% of patients then survive for a period longer than one year (O´Brien et al. 1996).

According to Attali-Soussay et al. (2001) clinical signs often encountered include exophthalmos (84%), conjunctival hyperaemia (40%), third eyelid prolapse (28%), exposure keratitis (20%) and anomalies of the fundus (20%). Barnet and Grimmes (1972) mention the occurrence of retinal detachment. When the tumour affects the nerve tissue, there may be progressive neurological signs (Andrew 1999). Radiographs often show osteolysis (Mason et al. 2001). Retrobulbar tumours may form metastases, for

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Table 1
Types of retrobulbar tumours in the dog (Hendrix and Gelatt 2000; Martin et al. 2000; O´Brien et al. 1996; Langham et al. 1971; Barnett et al. 1967)

Types of retrobulbar tumours
Actinic cell carcinoma
Adenocarcinoma – unspecified
Anaplastic astrocytoma
Carcinoma – unspecified
Chondrosarcoma
Fibrosarcoma
Leiomyoma
Lymphoma
Malignant melanoma
Malignant meningioma
Mast cell sarcoma
Meningioma
Myxofibrosarcoma
Nasal adenocarcinoma
Osteogenic sarcoma
Osteochondrosarcoma
Osteosarcoma
Rhabdomyosarcoma
Salivary gland adenocarcinoma
Sarcoma – unspecified
Squamous cell carcinoma

example, to the lung tissue with subsequent relevant clinical problems (Martin et al. 2000).

Resection of tumours is the treatment of choice. There is a better prognosis when the resection is accompanied by a partial or total orbitectomy (O´Brien et al. 1996). For the proper resection of retrobulbar tumours a lateral (Spiess and Wallin-Hakanson 1999) or modified orbitotomy (Gilger et al. 1994) is often necessary.

Case Report

A female Hovawart dog, three-year old and weighing 29 kg, vaccinated, was presented at the clinic. The owner reported problems with the right eye lasting for 3 months. These problems started as conjunctival hyperaemia and epiphora. A private veterinary surgeon diagnosed the dog with a follicular conjunctivitis and selected a combination of antibiotics and corticosteroids (bacitracin, neomycin, hydrocortisone) for the treatment. The finding on the eye was not getting better. On the contrary, it was getting worse and exophthalmos became evident during the last month of duration of the disease, in particular.

Apart from the affection of the right eye, the patient was without any health problems. Clinical examination and evaluation of the general state of health resulted in finding no other abnormalities. Upon ophthalmological examination we noted evident craniolateral exophthalmos with protrusion of the third eyelid and enlargement of the accessory lacrimal gland. There was a mild chemosis of the affected eye. There were no changes of size as well as shape of the globe as compared to the contralateral one (Plate VII, Figs 1, 2). No changes were found during a thorough examination (tonometry, slit lamp, ophthalmoscopy) we found any. The intraocular pressure (measured by Schiötz tonometer) was within physiological values (i.e., 21 mm Hg).

The patient was blood sampled for haematology and biochemical profile determination and sent to the department of diagnostic imaging. Here the patient was examined using radiography, sonography and computer tomography with special respect to the right orbit. No

abnormalities were found in the haematological and biochemical values. For the radiographic examination of the skull laterolateral and dorsoventral projections were taken. Neither of the projections showed signs of affection of the bony base. In the area of the right orbit there was only a mild shadowing of soft tissues. Computer tomography (Plate VII, Fig. 3) revealed a marked protrusion of the right globe due to the tumourous infiltration of extraocular muscles of the ventral group, in particular. Infiltrated muscles formed the tumourous infiltration of 3 cm in diameter completely filling the retrobulbar space. Cranially, a close intimacy of the infiltrate and the base of the skull was evident. It was not possible to rule out osteolysis without any doubts. Ultrasonographic examination of the globe and orbit resulted in finding a hyperechogenic mass of 10×8 mm in size in the caudomedial part of the orbit. It was clearly demarcated and did not penetrate into structures of the globe. There were no ultrasonographic abnormalities of the globe present. A fine needle biopsy was taken under sonographic guidance and the obtained sample was examined cytologically.

Cytology revealed cellular structures with an admixture of erythrocytes. There were individual cells, occasionally forming clusters, having cytoplasmic projections typical for the connective tissue. Sporadically, there was basophilia of the cytoplasm, marked anisocytosis and anisokaryosis, round to oval nuclei containing 1 to 6 nucleoli.

On the basis of these examinations we made the diagnosis of a malignant mesenchymal tumour and proposed its excision in order to make a definitive histological diagnosis.

General anaesthesia in the patient was induced using a combination of medetomidine and propofol and then maintained by inhalation of a mixture of oxygen, nitrous oxide and halothane. Following skin incision and preparation of subcutaneous tissues, lateral orbitotomy including arcus zygomaticus resection was performed (Plate VIII, Fig. 5). The retrobulbar space was accessed by retraction of m. rectus lateralis dorsally (Fig. 6). The tumour originating in m. rectus ventralis penetrated partly os presphenoidale and caused its osteolysis (which was not visible on the radiograph). The tumour was excised in its entirety, bleeding controlled by cauterisation and locally applied epinephrine. Arcus zygomaticus was then re-transposed and fixed by 4 bone sutures of surgical wire (Fig. 7). Fasciae, subcutaneous tissues and skin were sutured using polydioxanone 4/0, 3/0 and nylon 3/0, respectively. The operation lasted for 120 minutes and then the patient was given atipamezole. To reduce pain and local oedema, a single dose of flunixin meglumine was given to the dog. A combination of amoxicillin-clavulanate and enrofloxacin was used as an antibiotic shield. The resected tumour was sent for a histological evaluation. On day two following the operation there was a mild postoperation oedema in the area of the wound and of the conjunctiva of the lower eyelid. The globe was without any marked clinical changes. After two days the patient was released from the hospital. At the time of suture removal after 10 days, the eye was calm, clinically identical to the finding on the contralateral side and the patient showed no problems.

For histology the specimen was supplied in a formalin fixative. It was routinely processed for light microscopy and stained with hematoxylin-eosin and the PAS method. Immunohistochemical methods to confirm and type the intermediate filaments were necessary to reach the final diagnosis. The histological examination revealed a malignant, highly vascularised tumour consisting of polymorphic, mitotic-active cell populations (including atypical mitotic figures) and prevailing spindle-cell elements arranged mainly perivascularly. Immature myoblasts were frequent. Cytoplasmic cross-striation of some cells was observed. In the sample there were sporadic elements containing several nuclei or bizarre-shaped nuclei. Perivascular highly cellular areas alternated with areas of abundant intercellular mucoid material and extravasates with clumps of siderophagocytes (Fig. 8). The tumorous cells were positive for desmin and vimentin immunohistochemically.

Three months later, the female dog was brought at the clinic with a recurrence of the same extent and a similar clinical finding as during the first visit. Because of a poor prognosis euthanasia was recommended and performed with consent of the owner. The repeated histological examination confirmed recurrence of the rhabdomyosarcoma.

Discussion

Rhabdomyosarcoma is the type of a tumour in the human medicine reported as one of the most common soft tissue sarcomas in children and adolescents with a rather uncertain prognosis (Illanes 2002). In the dog it has been found in many organs. In the orbit, however, it has been reported only once. Of the many studies of this tumour, only Hendrix and Gelatt (2000) mention its association with the retrobulbar space.

Kim et al. (1996) reported two types of rhabdomyosarcomas, i.e., the alveolar and embryonal ones. Kuwamura et al. (1998) describe the rhabdomyosarcoma as a tissue formed by mature rhabdomyoblasts with an elongated eosinophilic cytoplasm and infrequent cross-striation. Tumourous cells are immunopositive for anti-myoglobin, desmin and vimentin antibodies. Ultrastructurally, they contain a quantity of myofibrils as well as Z-bands in the cytoplasm.

Occurrence of the rhabdomyosarcoma (bothryoid sarcoma) in the urinary bladder has relatively frequently been reported (Takiguchi et al. 2002; Kuwamura et al. 1998; Kim et al. 1996). Many authors reported occasional cases of finding the rhabdomyosarcoma in the heart atrium and chamber (Krotje et al. 1990; Gonin-Jmaa et al. 1996; Perez et al. 1998), perineal area (Ueno et al. 2002), parameningeal area (Illanes 2002), tongue (Lascelles et al. 1998), gingiva (Kim et al. 1996) or greater omentum (Sarnelli et al. 1994). Yanoff et al. (1996), Block et al. (1995) and Hendersen et al. (1991) report rhabdomyosarcomas causing tracheal defects.

Rhabdomyosarcomas are relatively frequently reported in purebred dogs of large breeds such as Labrador Retriever (Kuwamura et al. 1998; Illanes 2002; Ueno et al. 2002), Basset Hound (Kim et al. 1996), Rottweiler (Kim et al. 1996) or German Shepherd (Perez et al. 1998). In our case it was also a large breed dog – a Hovawart. Considering the age of affected animals, rhabdomyosarcomas are reported in young dogs of about 2 years (Yanoff et al. 1996; Takiguchi et al. 2002; Kuwamura et al. 1998; Kim et al. 1996; Illanes 2002). This fact corresponds to data from the human medicine on the occurrence of the rhabdomyosarcoma in children and adolescents (Illanes 2002). Data on the occurrence of the rhabdomyosarcoma in dogs of middle age are lacking. The second group of affected dogs is represented by older animals, mostly above 6 years (Block et al. 1995; Hendersen et al. 1991).

Due to the aggressiveness of the rhabdomyosarcoma, metastases to the liver (Illanes 2002) or regional lymph nodes and lungs are reported (Kim et al. 1996). In our case we did not perform aimed intravital diagnostic examinations (RTG, USG); autopsy, however, showed no metastases.

Because of a poor access to the tumour, the resection from the retrobulbar space was done using the technique of lateral orbitotomy and resection of arcus zygomaticus with its retraction dorsally (Spiess and Wallin-Hakanson 1999). Apart from this traditional technique, Gilger et al. (1994) described also a modification of lateral orbitotomy, which was not employed in this case due to the necessity of transection of lig. orbitale. Due to the fact that there were no radiographic signs of osteolysis, the tumour was resected without a concurrent partial or total orbitectomy, which would, with respect to the aggressive nature of the tumour, provide a better prognosis for the patient (O´Brien et al. 1996). Macroscopically, the tumour was resected in its entirety.

Recurrence of the same extent of rhabdomyosarcoma and similar clinical findings as prior to the surgery three months later is in agreement with references on recurrences of retrobulbar tumours reporting the survival of up to one year after the resection (Attali-Soussay et al. 2001; Hendrix and Gelatt 2000). The only study reporting more than 70% of patients living longer than one year is that of the use of orbitectomy (O´Brien et al. 1996) which, however, was not employed because of the above-mentioned reasons. Due to disagreement of the owner with chemotherapy - e.g., VAC protocol (Ueno et al. 2002), this type of therapy was not initiated.

Retrobulbární rabdomyosarkom u hovawarta

Klinický případ popisuje tříletou fenu hovawarta s jednostranným exoftalmem, u níž byl sonograficky a počítačovou tomografií diagnostikován retrobulbární tumour vycházející z m. rectus ventralis. Cytologicky byl tumor klasifikován jako maligní mezenchymální. Byla provedena modifikace laterální orbitotomie s kantotomií a makroskopicky ohraničený tumour byl exstirpován v celém jeho rozsahu. Po zhojení bylo oko ve stejném stavu jako oko zdravé, kontralaterální, po 3 měsících však nastala recidiva ve stejném rozsahu a pacient byl na žádost majitele utracen. Opakovaným histologickým vyšetřením byl potvrzen rabdomyosarkom.

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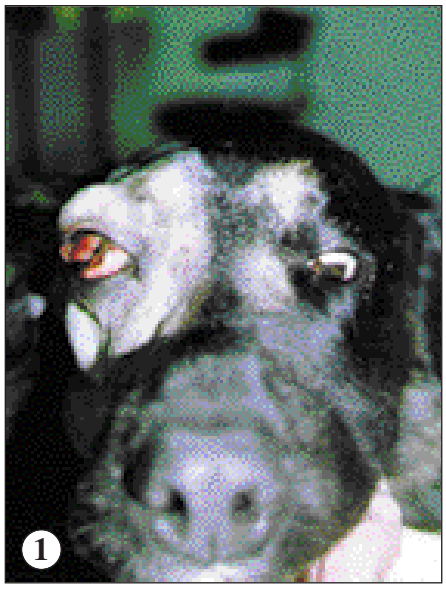

Fig. 1: Craniolateral exophthalmos, third eyelid prolapse and enlargement of the accessory lacrimal gland of the affected right eye. The globe, however, is of the same size as the healthy contralateral one.

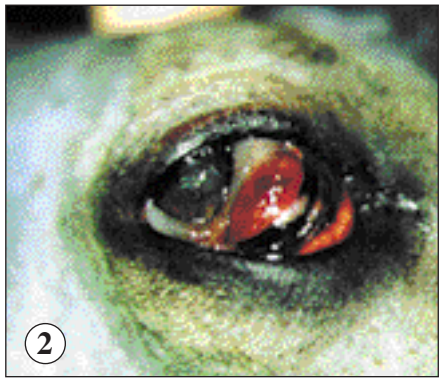

Fig. 2: The affected right globe in greater detail – there is hyperaemia, third eyelid prolapse and enlargement of the accessory lacrimal gland as well as conjunctival chemosis visible at the lateral canthus.

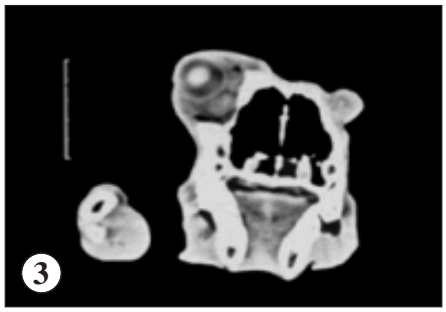

Fig. 3: A CT scan showing protrusion of the right globe due to the tumourous infiltration of ventral eyeball movement muscles in the size of about 3 cm filling completely the retrobulbar space. Osteolysis cannot be excluded without any doubts.

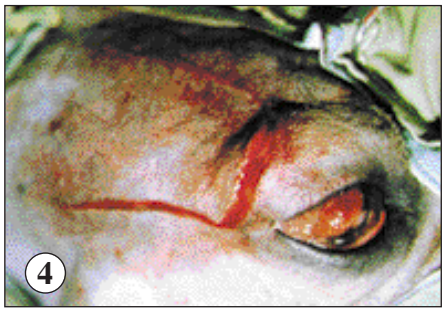

Fig. 4: A horizontal U-shaped skin incision during the lateral orbitotomy located dorsolaterally from the canthus along arcus zygomaticus.

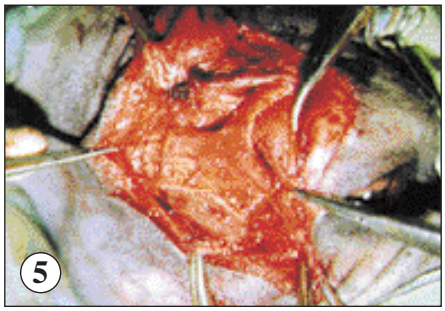

Fig. 5: Resection of arcus zygomaticus by vertical osteotomy cuts located cranial and caudal to the insertion of ligamentum orbitale following reflection of skin and subcutaneous tissues.

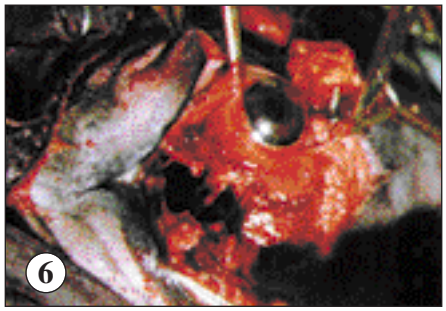

Fig. 6: A part of arcus zygomaticus and m. rectus lateralis drawn away dorsally in order to make access to the retrobulbar space and the tumour.

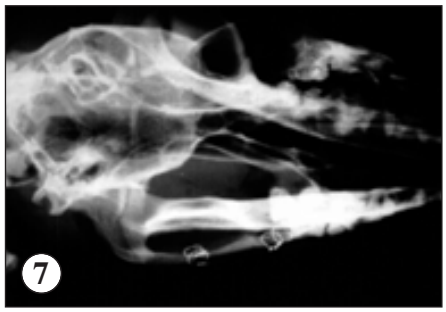

Fig. 7: A postoperation radiograph – bone sutures fixing the resected arcus zygomaticus. There is no osteolysis of os presphenoidale visible in the radiograph.

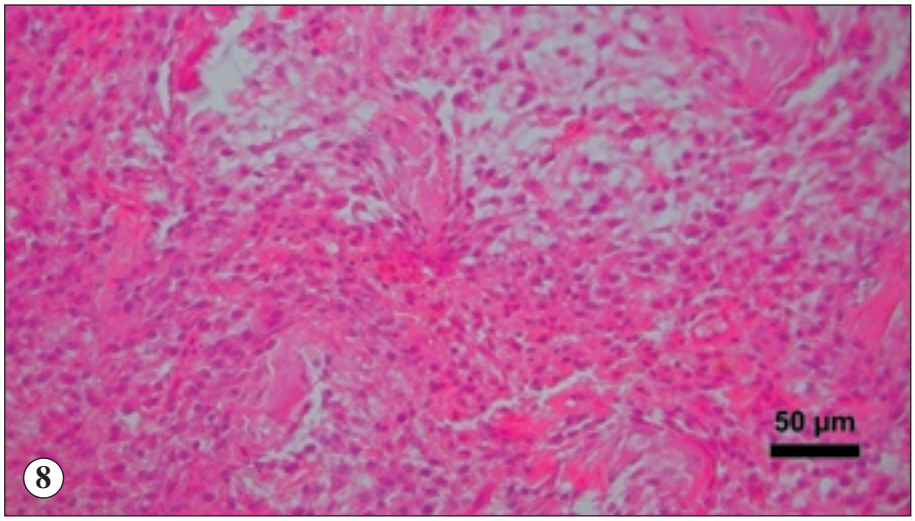

Fig. 8: A malignant tumour consisting of a population of polymorphic and mitotic-active cells with prevaling spindle-cell elements. There are relatively frequent immature myoblasts, sporadic elements with multiple nuclei, extensive extravasation and clumps of siderophagocytes (hematoxylin-eosin, magnification × 20).