Multiple congenital cardiac anomalies and idiopathic thoracic aortitis in a horse

G. P. Reppas, P. J. Canfield, W. J. Hartley, D. R. Hutchins, K. L. Hoffmann

An echocardiographical and clinical pathological investigation of the rapid loss of weight by a yearling thoroughbred filly revealed an atrial septal defect, a ventricular septal defect and hyperfibrinogenemia. A post mortem examination confirmed the cardiac abnormalities and revealed a severe thoracic aortitis. It is proposed that the idiopathic thoracic aortitis contributed to the horse's compromised cardiovascular homeostasis.

A WIDE range of congenital heart defects have been recognised in the horse but little is known about their relative frequency or causation. The most commonly reported abnormalities, in descending order of frequency, include ventricular septal defects, aortic stenosis, pulmonary stenosis, fenestration of the semi-lunar valves, persistent truncus arteriosus and ativoventricular valve defects (Rooney and Franks 1964, Whitney 1975, Huston and others 1977, Lombard and others 1983, Crowe and Swerczek 1985, Glazier 1986, Wilson and Haffner 1987, Leipoldt and others 1990, Ecke and others 1991). Ventricular septal defects may occur alone or in association with other defects (Hadiow and Ward 1980, Lombard and others 1983).

Acquired morphological changes in the aorta of the horse have been reported by Heidenreich (1960) and Maxie (1993). Many of these lesions exhibited proliferative and degenerative, rather than inflammatory, changes in the media and intima of the aorta (Imazumi and others 1989). Spontaneous rupture of the aorta has been the cause of the sudden death of some horses (Rooney and others 1967, Holmes and others 1973).

This report describes the clinical, echocardiographical and pathological findings in a horse with multiple congenital cardiac anomalies and thoracic aortic arteritis.

Case history

A bay yearling thoroughbred filly was examined because it had lost weight rapidly over a period of four weeks. Two months earlier the filly had been sold at auction and then transported and pad-docked with a small group of other fillies. The filly was inappetent on pasture and was reluctant to move about freely. When boxied it appeared to eat more satisfactorily.

The physical examination revealed that the horse's temperature (38.6°C) and respiration rate (20 breaths/min) were slightly raised, but its heart rate (48 beats/min) was normal. Upon auscultation of the thorax a loud 3/5 holosystolic murmur was audible on the left side and a 5/6 holosystolic murmur was audible on the right side. A prominent jugular pulse could be easily observed along both sides of the neck.

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<table>
<thead>
<tr>
<th>Cardiac parameter</th>
<th>Measurement</th>
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<tr>
<td>RVdD</td>
<td>4-52 cm</td>
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<tr>
<td>RVdS</td>
<td>3-2 cm</td>
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<tr>
<td>IVsD</td>
<td>3-46 cm</td>
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<tr>
<td>IVsS</td>
<td>4-29 cm</td>
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<tr>
<td>LVIDD</td>
<td>11-6 cm</td>
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<td>LVIDS</td>
<td>7-63 cm</td>
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<td>LVPWd</td>
<td>2-27 cm</td>
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<tr>
<td>LVPWb</td>
<td>3-88 cm</td>
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<tr>
<td>HR</td>
<td>52 bpm</td>
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<tr>
<td>FS</td>
<td>34-2 per cent</td>
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<tr>
<td>ET</td>
<td>0-36 s</td>
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<tr>
<td>AoD</td>
<td>7-48 cm</td>
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<tr>
<td>LAD</td>
<td>11-5 cm</td>
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<tr>
<td>PAD</td>
<td>7-13 cm</td>
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AoD Ascending aorta from the right parasternal long axis view, ET Ejection time, FS Per cent fractional shortening, HR Heart rate, IVsD Intraventricular septal thickness in diastole, IVsS Intraventricular septal thickness in systole, LAD Left arterial diameter from the left parasternal long axis view, LVIDD Left ventricular internal diameter in diastole, LVIDS Left ventricular internal diameter in systole, LVPWd Left ventricular posterior wall in diastole, LVPWs Left ventricular posterior wall in systole, PAd Pulmonary artery diameter from the left parasternal cranial angled view, RVdD Right ventricular diameter in diastole, RVdS Right ventricular diameter in systole

Routine haematological evaluations over three weeks revealed a mild, microcytic normochromic anaemia on two occasions. The animal also had a persistent hyperproteininaemia due to a mild to moderate hyperglobulinaemia. A marked hyperfibrinogenemia (10 g/litre) was recorded four days before the animal was euthanased. Abdominal paracentesis revealed no abnormalities and thoracic radiographs were within normal limits. Blood culture was negative.

Echocardiography

Echocardiography revealed multiple cardiac anomalies. The abnormalities identified included a 4 cm ventricular septal defect, a 6 cm ostium secundum atrial septal defect and an overriding aorta. The aortic ejection time was short (0-36 seconds). There was an enlargement of the right side of the heart with a prominent moderator band (septomarginal trabecula). The septal motion was flattened during diastole in both B and M mode imaging (Table 1).

Pathology

Tissue samples, fixed in 10 per cent buffered formalin were paraffin-embedded, sectioned at 5 to 6 μm, and stained with haematoxylin and eosin.

The animal was in poor condition with evidence of muscle wasting. Both lungs were congested and mildly oedematous. The trachea was filled with fine white foam which extended down into the smaller airways. The liver was slightly shrunken and red-brown. There was subcutaneous oedema of the extremities of all the limbs.

On examination of the heart there was a 6 cm diameter atrial septal defect and a high, subaortic, 4 cm diameter, ventricular septal defect. The right atrium was thin walled and markedly dis tended. Both papillary muscles in the right ventricle were hyper trophyed. At the base of the papillary muscle in the right ventricle lying closest to the interventricular septum there was a discrete, well circumscribed area of pallor. The chordae tendinae arising
from the papillary muscle lying farthest away from the interventricular septum were shortened. The moderator band (septomarginal trabecula) in the right ventricle was 1 cm in diameter. There was overriding of the right ventricle by the aorta.

Within the lumen of the thoracic aorta, beginning about 20 cm from its origin at the base of the heart, was a 15 cm long, circumferential, transmural, dull-grey, non-ulcerated, raised lesion which was 1-5 cm thick (Fig 1). A cross-section revealed a plaque of firm grey tissue overlying an area of brown-red discoloration.

Histopathological examination of the thoracic aortic lesion revealed a large intimal focus of neutrophils surrounded by dense fibrous tissue and an admixture of lymphocytes, plasma cells and macrophages (Fig 3). Using a Perl’s stain some of the macrophages contained haemosiderin. No bacteria were identified by a Brown and Brenn stain in the intimal plaque.

In sections of tunica media stained by the Elastica/van Gieson’s procedure, there were several irregularly shaped, poorly demarcated areas showing a loss and disruption of elastic fibres and smooth muscle (Fig 2). There was a moderate increase in the amount of collagen in these areas. Necrosis and calcification of elastic fibres were demonstrated by the use of Von Kossa’s stain. These areas were distributed throughout the width of the tunica media and subjacent to the intimal lesions.

Microscopical examination of the small focus of myocardial pallor at the base of the papillary muscle in the right ventricle revealed myocardial necrosis with evidence of replacement fibrosis. Several other tissues were examined but no other lesions were found.

Discussion

The rapid loss of weight by the yearling thoroughbred filly shortly after its sale at auction prompted a thorough medical investigation of its condition. Although an echocardiographical and clinical pathological examination revealed an atrial septal defect, a ventricular septal defect and hyperfibrinogenemia, the severe thoracic aortitis was found only post mortem.

In the past, a confirmation of the clinical diagnosis of ventricular septal defects in large animals has depended on cardiac catheterisation studies or necropsy. With the advent of echocardiography, ante mortem diagnoses of such conditions have become routine (Pipers and others 1985). Ventricular septal defects usually result in the shunting of blood from the left ventricle to the right ventricle, with concurrent pulmonary overcirculation. Affected animals are often unthrifty, die prematurely or are euthanased at an early age (Lombard and others 1983, Pipers and others 1985, Ecke and others 1991). Occasionally horses with a large ventricular septal defect have been reported to have remained clinically normal until one to two years of age, when the cardiovascular demands increase (Lombard and others 1983, Pipers and others 1985). More commonly, horses with isolated, small (<2-5 cm diameter), restrictive membranous ventricular septal defects without signs of volume overload and with left to right flow (>4 m/s) have been able to perform ( Reef 1991).

Few cases have been reported in which a horse has had a combined ventricular septal defect and atrial septal defect (Pipers and others 1985, Ecke and others 1991). The two cases described were both less than one month of age, had cyanotic mucous membranes, and died shortly after examination. The fact that this filly maintained its condition until the yearling sales, despite having both defects, was unusual.

Excessive thickening of the moderator bands (trabecula septomarginalis) in the right ventricle bridging the ventricular septum and free wall is uncommon in horses (Wilson and Haffner 1987). It may accompany hypertrophy of the right ventricle, when there is increased thickness of the wall of the affected chamber and a remarkable increase in the size of the papillary muscle and trabeculae carnae (Maxie 1993).

In this horse a small, 1 cm diameter, well circumscribed focus of myocardial necrosis with replacement fibrosis was evident at the base of the papillary muscle in the right ventricle. Hearts that are hypertrophic are extraordinarily susceptible to patchy myocardial necrosis of apparently random distribution, although large lesions are more obvious in the papillary muscles of the left ventricle. Replacement fibrosis is usually well established by the end of the sixth week after the initial insult to the myocardium (Maxie 1993).

The moderate to marked increase in fibrinogen concentration in this filly was one of the few clinical pathological aberrations observed. Fibrinogen is probably the most commonly measured acute phase protein in animals (Schalm and others 1970), although it is not the major acute phase protein in animal species (Laurel 1985), because it is easily measured in plasma (Benjamin 1978). Acute phase proteins such as fibrinogen, are a group of mainly glycoproteins produced by the liver in response to tissue damage. The level of response is ideally equivalent to the amount of tissue damaged. Plasma fibrinogen levels in the absence of dehydration are increased in a wide variety of inflammatory and tissue destructive conditions, including degenerative, traumatic and neoplastic...
diseases (Jain 1986). The possibility of a significant septic focus associated with the heart valves, and accompanying this filly’s congenital cardiac defects, was considered, but the thorough post mortem examination failed to find one. However, the examination did suggest an alternative source of inflammation and tissue destruction as a focus which could account for the horse’s hyperfibrinogenaemia.

Lesions in the aorta have been observed during investigations of the morphological changes in the great vessels of horses (Heidenreich 1960, Cranley 1983, Imaizumi and others 1989). These studies revealed a high incidence of degenerative lesions in the wall of the aorta, especially in horses with a racing career. In some studies inflammatory lesions of the aorta were not commonly found (Imaizumi and others 1989). The lesion in this horse’s thoracic aorta was diagnosed histopathologically as focal chronic supplicative aortitis, the aetiology of which could not be determined from the material examined.

The thickening of the thoracic aorta and the proliferative intimal lesions were attributable to fibroplasia and a mixed inflammatory cell infiltrate comprising neutrophils, lymphocytes and plasma cells (Fig 3). The lack of eosinophils in the affected area, the absence of endoarteritis in the cranial mesenteric artery and a history of regular worming since weaning tended to discount, but could not completely eliminate, the possibility of a parasitic aortitis due to Strongylus vulgaris infection. Although one study has suggested that the larvae of *S. vulgaris* are unlikely to migrate to the aorta after passing through the heart and lungs (Imaizumi and others 1989), other reports suggest that aberrant larvae do migrate through the walls of the thoracic and proximal abdominal aorta (Farrell 1954, Cranley and McCullagh 1981). Intimal tracts, endoarteritis with thrombosis and fibrous nodules of parietal origin are sometimes encountered in the bulb of the aorta, usually on its cranial curvature and in the region of the coronary sinuses. A detailed histopathological examination of various organs failed to substantiate the involvement of arteries other than the aorta in this horse. Most viral arteritides in the horse are widely disseminated throughout the body, for example, equine viral rhinopneumonitis/encephalomyelitis. Other causes of focal vasculitis associated with partial occlusive disease of the thoracic aorta include the deposition of immune complexes as well as bacterial agents (Maxie 1993).

In human beings inflammatory lesions of the arteries are uncommon. Septic emboli or bacteraemia usually account for arterial infections at a considerable distance from their source, with the vasa vasoem being the route of entry into large vessels such as the aorta. The direct entry of organisms from the surface occurs only in vessels small enough to entrap infectious particles or bacterial clusters. In the very rare instances of primary acute aortitis in human beings, the source of the infection – either endocarditis, or periarterial or arterial inflammation – cannot be demonstrated (Gore 1968).

The filly was in reasonable condition during the yearling sale and it is suggested that the rapid deterioration in its demeanour and condition may have been the result of a fulminant, partially occlusive, inflammatory thoracic aortic lesion which destabilised the horse’s compromised cardiovascular homeostasis.

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