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Brief Report

Neonatal aortic arch obstruction due to pedunculated left ventricular foetal myxoma

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Abstract Myxoma in neonatal life are extremely rare. We report a case of a neonate with a pedunculated cardiac tumour arising from the anterolateral left ventricular wall protruding across the left ventricular outflow tract and continuously extending into the distal aortic arch. Surgical removal at 14 days of age via combined transaortic approach and apical ventriculotomy was indicated because of the risk of further compromise of aortic valve function and aortic arch obstruction. Histopathologic examination was consistent with a myxoma.

Keywords: Myxoma; cardiac tumour; left ventricular outflow tract obstruction; foetal studies

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Case report

Primary cardiac tumours during foetal life and infancy are a rare finding described with an incidence of 0.14%.[1] Most of the foetal und neonatal cardiac tumours are benign; haemodynamic relevance and clinical symptoms will depend on size, number, and localisation of the tumour. During foetal life, pericardial and pleural effusion, cardiac dysfunction or hydrops, and arrhythmia or conduction disturbances might develop, and postnatally inflow or outflow obstruction will determine the clinical symptoms.[1,2] Rhabdomyoma and fibroma are the most common primary heart tumours in infancy. Myxomas are more frequent in older children predominantly arising at atrial level.[1,3] Until 2003, myxomas have never been described in a foetus,[4] and only a few reports describe myxoma in the neonatal period arising from the right atrial and right ventricular structures.[5,6]

On the first day of life, the neonate was transferred to our unit because of the prescribed foetal ultrasound finding from the 25-week examination with a mild left ventricular outflow tract obstruction caused by a mass protruding across the aortic valve annulus in the presence of an otherwise normally developed left ventricle, aortic valve annulus, and aortic arch. On postnatal transthoracic echocardiography, an echogenic, pedunculated tumour was demonstrated arising at the anterolateral left ventricular wall next to the anterior papillary muscle extending across the left ventricular outflow tract and partially obstructing the structurally normal aortic valve (Fig 1a). There was no significant gradient across the outflow tract and only an insignificant aortic insufficiency. The tumour was loosely attached at the sinutubular junction with continuous extension into the aortic arch along its inferior curvature and isthmic region, with some adhesions superior and inferior to the origin of the left subclavian artery (Fig 1b). On colour flow mapping, there was some flow disturbance, and maximum blood flow velocity within the distal arch to the descending aorta was increased to 2.5 m/second. The infant was asymptomatic. Criteria for surgical removal at the age of 14 days were the risk for thromboembolic complications, further compromise of aortic valve function, and aortic arch obstruction. The intracavitary tumour was removed through an apical left ventriculotomy using cardiopulmonary bypass and hypothermic circulatory arrest. This approach allowed for detailed exposure and complete resection of the tumour, inserting nearby the anterolateral papillary muscle. The transaortic approach was used for removal of the transvalvular and intraluminal extension of the tumour within the aortic arch by careful suction. The postoperative period was uneventful.

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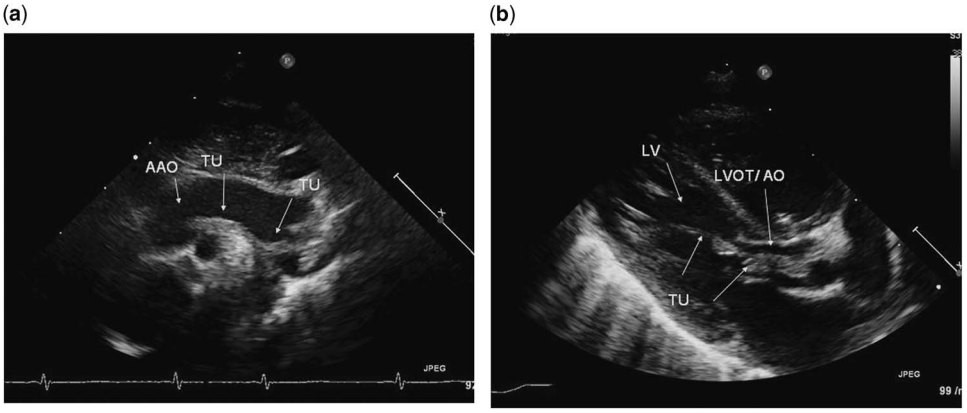

Figure 1.
*(**a**) Parasternal long-axis view with longitudinal extension of the left ventricular tumour across the left ventricular outflow tract. (**b**) Aortic arch view at the level of origin of the left subclavian artery: tumour extension within the aortic arch and adhesion around the origin of the left subclavian artery (arrow). AAO = ascending aorta; AO = aorta; LV = left ventricle; LVOT = left ventricular outflow tract; TU = tumour.*

Histopathologic examination revealed ovoid cells with eosinophilic cytoplasma singly orientated and widely spaced in the myxoid stroma; elastic fibres and collagen bundles indicated fibrosis (Fig 2).

Myxomas, as described in our neonatal patient and confirmed on histopathological examination, are extremely rare lesions, especially with regard to left ventricular wall origin. The exceptional tumour extension from the left ventricular cavity through the left ventricular outflow tract into the aortic arch has not been observed yet. Myxomas account for <1% of cardiac tumours in paediatric patients; they are more often found in older children and adult patients.[1] The tumour appears as an echogenic, pedunculated, or broadly attached structure mainly localised in the left or right atrium. In children, a much more variable site of the origin of the myxoma has been considered, including the right ventricular wall or infundibulum.[6,7] Originating from the right or, extremely rare, left ventricular endocardium clinical symptoms of inflow or outflow obstruction have been described in adult patients.[8] In this situation, protrusion of the mass through the aortic valve caused severe acute left ventricular outflow tract obstruction; the risk of sudden death has to be considered, and urgent removal of the mass is indicated.[8]

Tumour localisation and unusual tumour extension in our neonate might be explained by further extension of the tumour mass during pregnancy guided by the direction and streaming of blood flow. Postnatal echocardiography demonstrated extension of the echogenic mass within the aortic arch and some adhesions at the origin of the left subclavian artery – opposite to the entrance and predominant direction of streaming of the foetal arterial duct. An additional explanation for flow-related tumour extension might be that myxoma are thought to

Figure 2.
Histopathologic examination revealed the ovoid cells with eosinophilic cytoplasma singly orientated and widely spaced in the myxoid stroma; elastic fibres and collagen bundles indicate fibrosis (alcian blue staining).

arise from the pluripotential mesenchymal cells dispersed in a myxoid stroma and have a soft, gelatinous consistency.[1,7]

Prenatal diagnosis of cardiac tumours have been described as early as 20 weeks of gestation mainly between 22 and 34 weeks; with further development during pregnancy the detection rate will increase. This observation has been made for rhabdomyomas and teratomas; fibroma or haemangioma have rarely been observed during infancy. Until 2003, myxoma have not been reported in a foetus and remain an exceptional finding on prenatal ultrasound.[3,4] Neonatal myxoma with severe symptoms of right atrial obstruction was reported in 1982.[9] George et al 2006 described a myxoma in a neonate arising from the

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infundibulum and partially obstructing the right ventricular outflow tract and found two other cases with right ventricular myxoma in the literature. Pathology studies in these cases were consistent with myxoma describing the stellate or ovoid cells with decreased or lack of mitotic activity within a myxoid matrix that undergoes fibrosis or calcification.[6,10] Histiogenesis of myxoma was described as "uncertain". Immunohistochemical study indicated that they arise from the pluripotential mesenchymal cells;[7] another assumption is that the stroma cells derived from the endocardial neural tissue.[10] The principle diagnostic criteria of the eosinophilc cells – scattered stallate/spindle cells, singly or organised as nests or chords in a myxoid stroma were also found in our patient. Treatment of choice is complete resection, as haemodynamic compromise, arrhythmia, or embolic events might develop.[5,6] Risk for recurrence is only given after incomplete resection or recurrence at multiple sites.

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Conflicts of Interest

None.

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