

## Pericardial Thymoma: An Unusual Cause of Sudden Death<sup>†</sup>

Dear Editor,

Thymomas are located in the normal location of the thymus, the anterior mediastinum. It is very rare for thymomas to arise primarily intrapericardially.<sup>1,2</sup> A survey of the literature reveals that only a few reports on these tumours are available.<sup>1,3</sup> While the most common complication of orthotopic thymoma is myasthenia gravis a paraneoplastic syndrome, patients with pericardial tumour manifestations often present with symptoms of impeded cardiac function.<sup>4</sup> We present a case of thymoma involving the pericardium. The tumour was an entirely intrapericardial thymoma.

According to the death certificate, the patient had lost consciousness during the night and was taken to a hospital emergency department where she was treated. However, she died on the same day. The death was considered of a suspicious nature and an autopsy was mandated by the prosecutor. Her family members reported that she had had occasional episodes of chest pain, fatigue, and that she had been diagnosed with a postpartum anxiety disorder. The primary location of the thymoma had not been clearly established by postpartum radiograms, and no abnormal mediastinal profiles had been noted. Analgesic treatment had been administered. She had no myasthenia-like symptoms, weight loss, night sweats, pruritus or lymphadenopathy. The patient was 23 years old, 169 cm in height and weighed 60 kg. On investigation, defibrillator marks on the thoracic wall and needle puncture sites on the back of the right hand and the cubital fossas were detected. The autopsy revealed a massively thickened pericardium, and a huge mediastinal mass. The tumour was encapsulated measuring 10 x 4 x 3 cm (Fig. 1). It showed a lobulated pink-tan cut surface. Areas of necrosis and haemorrhage were also noted. Macroscopic examination of both lungs revealed subpleural superficial bleeding areas between the lobes; histopathological examination showed oedema and congestion. The brain appeared normal on histopathological examination. The heart weighed 320 g. Tissues had been routinely fixed in 10% buffered formaldehyde, embedded in paraffin and subsequently stained with haematoxylin-eosin. Histologic sections showed a cellular tumour with a prominent lobular pattern under low-power microscopy. The lesions showed dense lymphoid tissue with a lobular configuration, sharply separated by thick collagenous septa arising from the capsule. With higher magnification, polygonal epithelioid cells admixed with

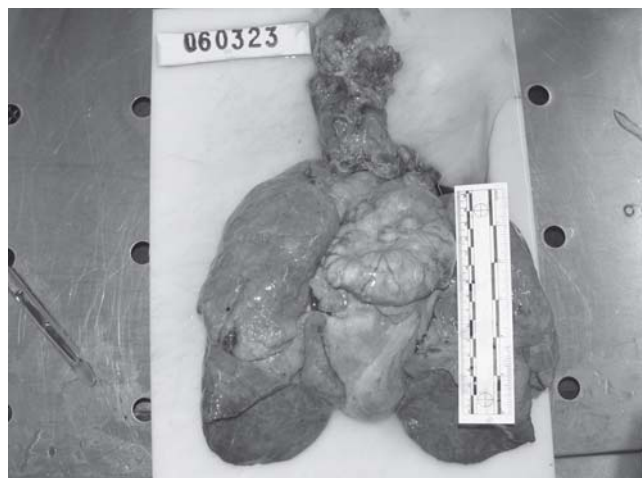


Fig. 1. Lobulated pericardial mass on autopsy examination.

the lymphoid tissue were visible. A panel of antibodies was applied to paraffin sections using commercially available reagents. The pale cells were positive for pancytokeratin, cytokeratin 7, and p63. Lymphoid cells were positive for terminal deoxynucleotidyl transferase, CD1A and CD5. A diagnosis of lymphositic thymoma was made. There was adjacent pleural thickening but there was no evidence of capsular penetration by the tumour. Ten sections of the heart were evaluated. Histopathological examination did not reveal evidence of acute or chronic ischaemia, but mild hypertrophy was recorded. Analysis of the organ specimens revealed none of the substances screened for in systematic toxicological methods. In this study, we present a case of thymoma involving the pericardium. The tumour was an entirely intrapericardial thymoma. The death was reported as sudden natural death due to mechanical heart compression by pericardial thymoma.

Pericardial involvement by thymic tumours is usually a late stage observation in primary mediastinal thymomas.<sup>1,2</sup> The development of massive pericardial effusion associated with pericardial metastasis of thymoma has been described by a number of authors. In some cases it was the first clinical sign of the thymic tumour.<sup>1</sup> Thymomas originating primarily in the pericardium, however, are rarely reported neoplasms. A review of the literature reveals a small number of cases of primary intrapericardial thymoma.<sup>1-3</sup> Intrapericardial thymoma may raise several considerations as far as its clinical and pathologic features are concerned. It is mainly observed in adult or elderly patients. On the

<sup>†</sup> This study was done with permission of Council of Forensic Medicine of Turkey Scientific- Ethic Commission

other hand, its symptom may be more evident than those of classical thymoma.<sup>5</sup> The histological subtype of primary intrapericardial thymoma was the spindle cell or medullary variant, classified by Marino and Müller-Hermelink.<sup>2</sup> However, the presenting case was lymphositic thymoma. The presence of mediastinal thymoma was excluded by autopsy establishing the diagnosis of a primarily intrapericardial thymoma. The primary pericardial location of the thymoma was not clearly established by radiograms in our case, but the pericardial thymoma determined the clinical course of the patient. Unlike our case, the patients were mostly elderly women who presented clinically with symptoms of right heart congestion.<sup>3</sup> It is likely that symptoms of intrapericardial tumour growth due to cardiac compression occur earlier than other thymoma-related complications, especially myasthenia gravis.<sup>4</sup> The pericardium has limited growth space. Symptoms caused by direct mechanical compression or pericardial effusion are therefore likely to occur early and present as the first clinical signs of the tumour.<sup>4,5</sup> Since the pericardium does not have the potential for growth that the anterior mediastinum does, shortness of breath, cough or chest pain are likely to develop earlier, even if the tumour has slow or stationary growth.<sup>3,4</sup> On the other hand, as in our case, pressure effect symptoms are non-specific and may be misinterpreted or attributed to other, often concomitant diseases of the elderly. Functional signs may be present, as they have been described in other ectopic thymomas, but they are uncommon for myasthenia gravis.<sup>5</sup> The origin of intrapericardial and other ectopic thymomas has been the subject of several speculative theories. Still, an abnormal development of the thymus gland during its phase of embryonic descent from the thymic anlage, associated with the misplacement of gland remnants, appears to be the most valid theory, as explained by Marino and Müller-Hermelink.<sup>2</sup>

This case study confirms that the pericardium is a possible rare location of thymic tumour and that pericardial involvement by thymomas should be considered in cases with episodes of chest pain and fatigue.

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