

Thymic teratoma presenting as non-immune hydrops fetalis

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ABSTRACT

Teratomas are one of the most frequent tumors in the pediatric population. They occur anywhere along the midline of the body, following the course of the embryonic germ cell ridge. In the mediastinal location, they exert space occupying effects, leading to a myriad of complications, including non-immune hydrops fetalis. We describe a fatal case of an immature thymic teratoma in a neonate presenting with hydrops fetalis. This case emphasizes the importance of early diagnosis and surgical intervention in such cases.

Keywords

Erythroblastosis fetalis, Hydrops fetalis; Mediastinal Neoplasms; Teratoma

INTRODUCTION

Hydrops fetalis is the collection of excessive fluid in fetal cavities and soft tissue. Non-immune hydrops fetalis (NIHF) comprises the subset that is not caused by red cell alloimmunization and accounts for almost 90% cases.^{1,2} The prevalence of NIHF ranges from 1/1500 to 1/4000 births.² The basic underlying mechanism of NIHF is dysregulation of fluid movement between the vascular and interstitial spaces. Cardiac abnormalities are the most common cause of hydrops fetalis.^{3,4} Among numerous other etiologies, thoracic abnormalities account for 6% cases^{3,4} and include congenital malformations and tumors causing impaired venous return. Among tumors, mediastinal/thymic teratoma comprise a substantial proportion of cases.

Teratomas are one of the most frequent tumors in the pediatric population.⁵ They are derived from all three embryonic layers and can be gonadal as well as extragonadal. Among extragonadal locations sacrococcygeal teratomas account for 40% while

mediastinal cases account for less than 10%.⁵⁻⁷ Mediastinal teratomas are usually located in the anterior mediastinum where they arise either from the thymus or ectopic thyroid tissue.^{5,6} We describe the autopsy findings in a 7-day old neonate presenting with non-immune hydrops fetalis secondary to mass effect from an immature teratoma of thymic origin.

CASE REPORT

A male infant was born at 31 weeks' gestation via caesarean section to a 33-year-old, G4P2 mother with hypothyroidism. The mother was compliant with all prenatal care and had an unremarkable gestation. Prior to presentation, she noted decreased fetal movements and ultrasonography demonstrated fetal hydrops, decreased fetal movement, and decelerations. Upon delivery, APGAR scores were 3, 6, and 7 at 1, 5, and 10 minutes, respectively. The baby weighed 2494 grams, had a

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crown-heel length of 43.9 cm and was noted to be hydropic (Figure 1), hypotensive, and in respiratory distress. Imaging demonstrated bilateral pleural effusions, atelectasis, pneumothorax and abundant ascitic fluid. A prominent thymus and an expanded cardiothymic silhouette were noted on chest radiograph (Figure 2).

The findings were worrisome for a mediastinal mass; however, the patient was deemed unstable for CT/MRI or any further invasive intervention. A 2D echocardiogram demonstrated pericardial effusion, a patent ductus arteriosus, mild tricuspid regurgitation, and trivial mitral regurgitation. Pericardiocentesis was performed and the newborn was treated with dopamine, furosemide, and albumin. Infectious disease was consulted to rule out infectious causes of hydrops fetalis; however, cultures and serology failed to yield any causative agents. Complete blood counts showed anemia and leukopenia, while arterial blood gases revealed marked acidosis. Despite aggressive medical management with high-flow oxygen, surfactant, and broad spectrum antibiotics, the infant died on seventh day of life and a post-mortem examination limited to the abdomen and thorax was performed.

AUTOPSY FINDINGS

The most significant finding at autopsy was an 84 gram, tan-brown mass arising from the left inferior horn of the thymus causing a left lateral shift of the heart and compression of the left lung with gross extension into the pericardial sac (Figure 3A and 3B).

The pericardial sac, bilateral pleural cavities, and abdomen contained abundant blood-tinged serous fluid. The right and left lungs weighed 27.5 and 17.53 grams, respectively. The left lung exhibited markedly decreased size and compression by the laterally displaced heart, which weighed 9.95 g and was structurally normal. On sectioning, the cut surfaces of both lungs demonstrated poor aeration. The pulmonary veins were dilated and encased by the thymic mass. The bilateral lungs demonstrated early signs of hyaline membrane disease, atelectasis, and poor alveolar expansion (Figure 4).

On histologic examination, the thymic mass was determined to be an immature teratoma with abundant immature neural elements (Figure 5 and 6).

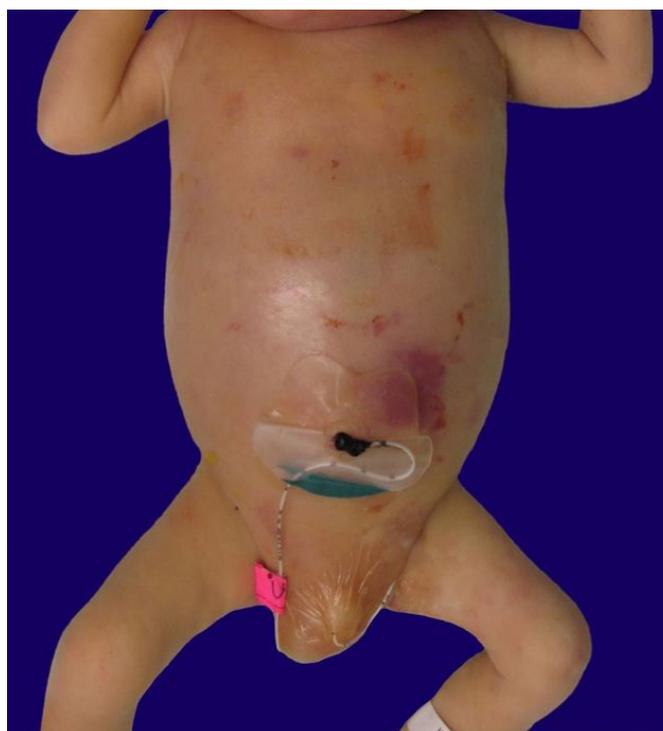


Figure 1. Generalized hydropic appearance.

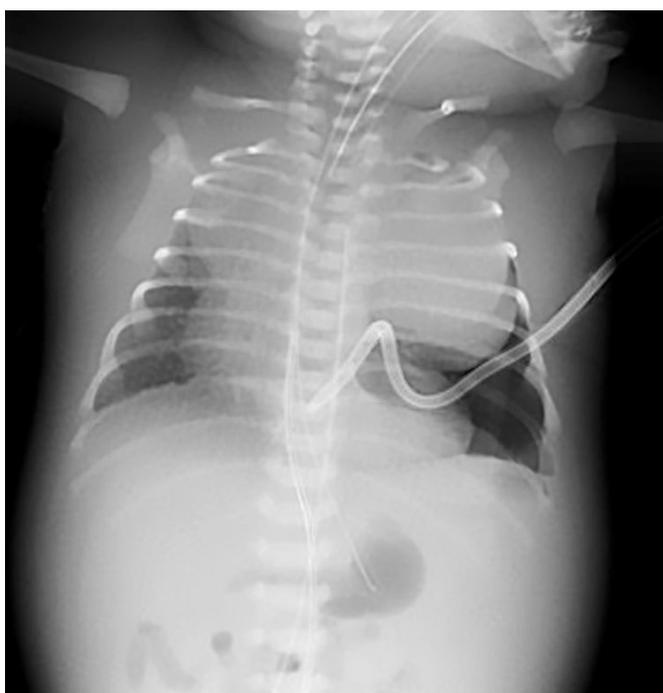


Figure 2. Chest x-ray showing expanded cardiothymic silhouette and persistent pneumothorax despite chest tube insertion.

The heart showed normal histology and the remaining organs displayed signs of vascular congestion. Table 1 demonstrates the weights of the infant's organs in comparison to the standard weights at 31 and 32 weeks of gestation.

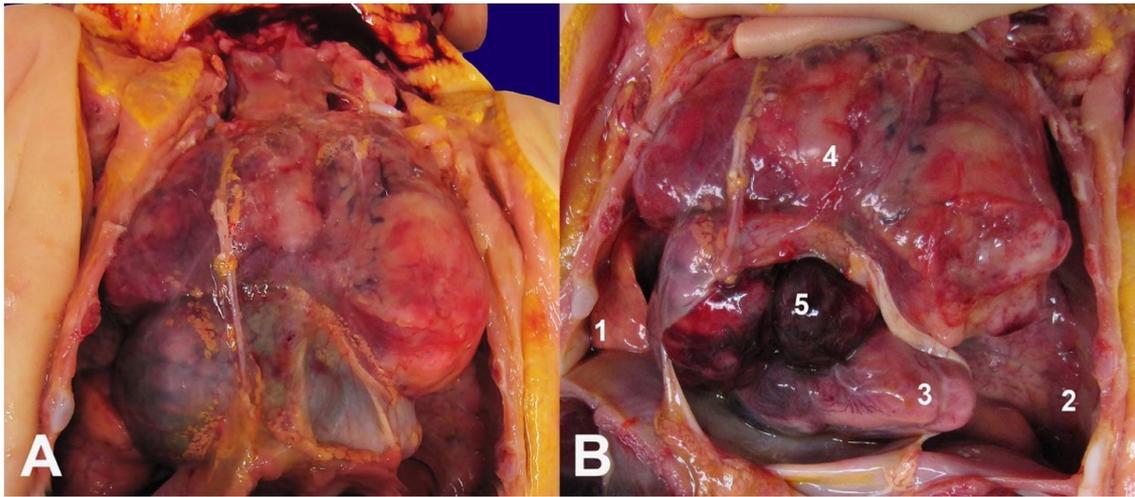


Figure 3. In-situ gross appearance of the thymic mass (**A** – Mass within intact pericardial sac; **B** – Pericardial sac opened to reveal in growing mass (1-Right lung; 2-Left lung; 3-Heart; 4-Thymic mass; 5-Mass extending into the pericardial sac).

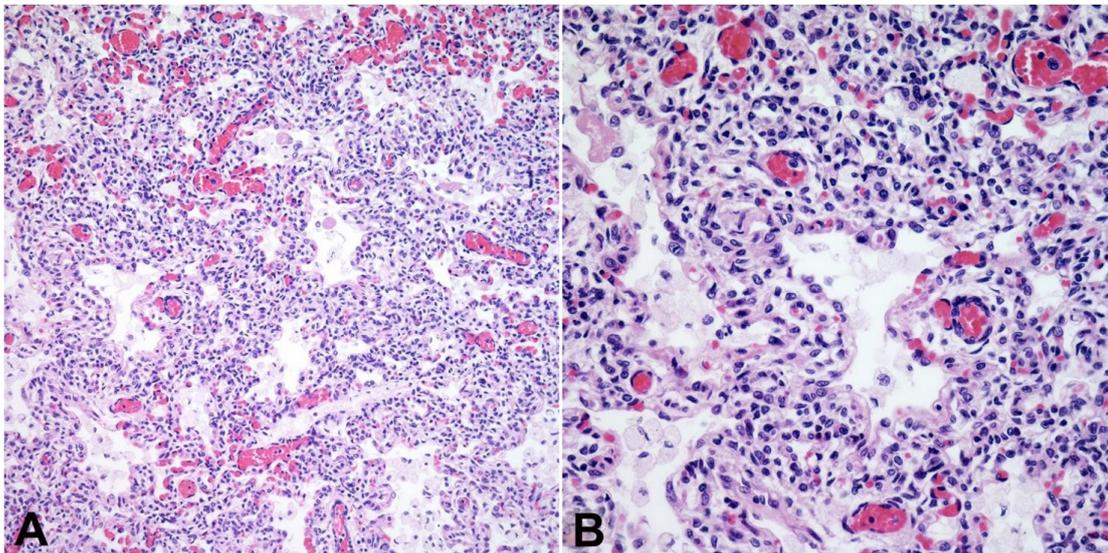


Figure 4. Photomicrograph of lungs demonstrating early signs of hyaline membrane disease, and poor alveolar expansion (**A** – H&E, 200X; **B** – H&E, 400X).

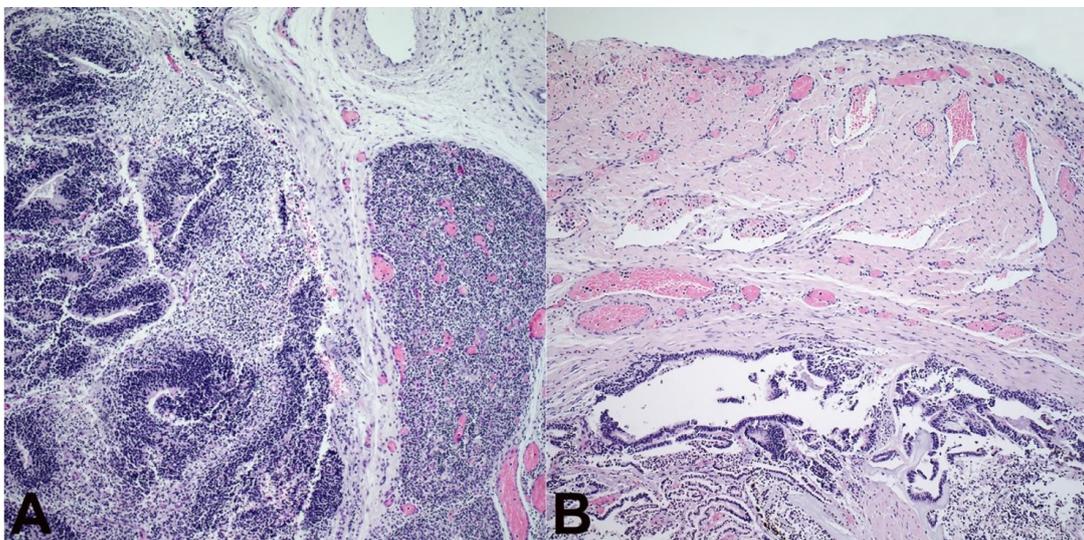


Figure 5. Immature neural elements of the teratoma with adjacent normal appearing thymic tissue (**A** – H&E, 100X). Teratoma extending into the pericardium (**B** – H&E, 100X).

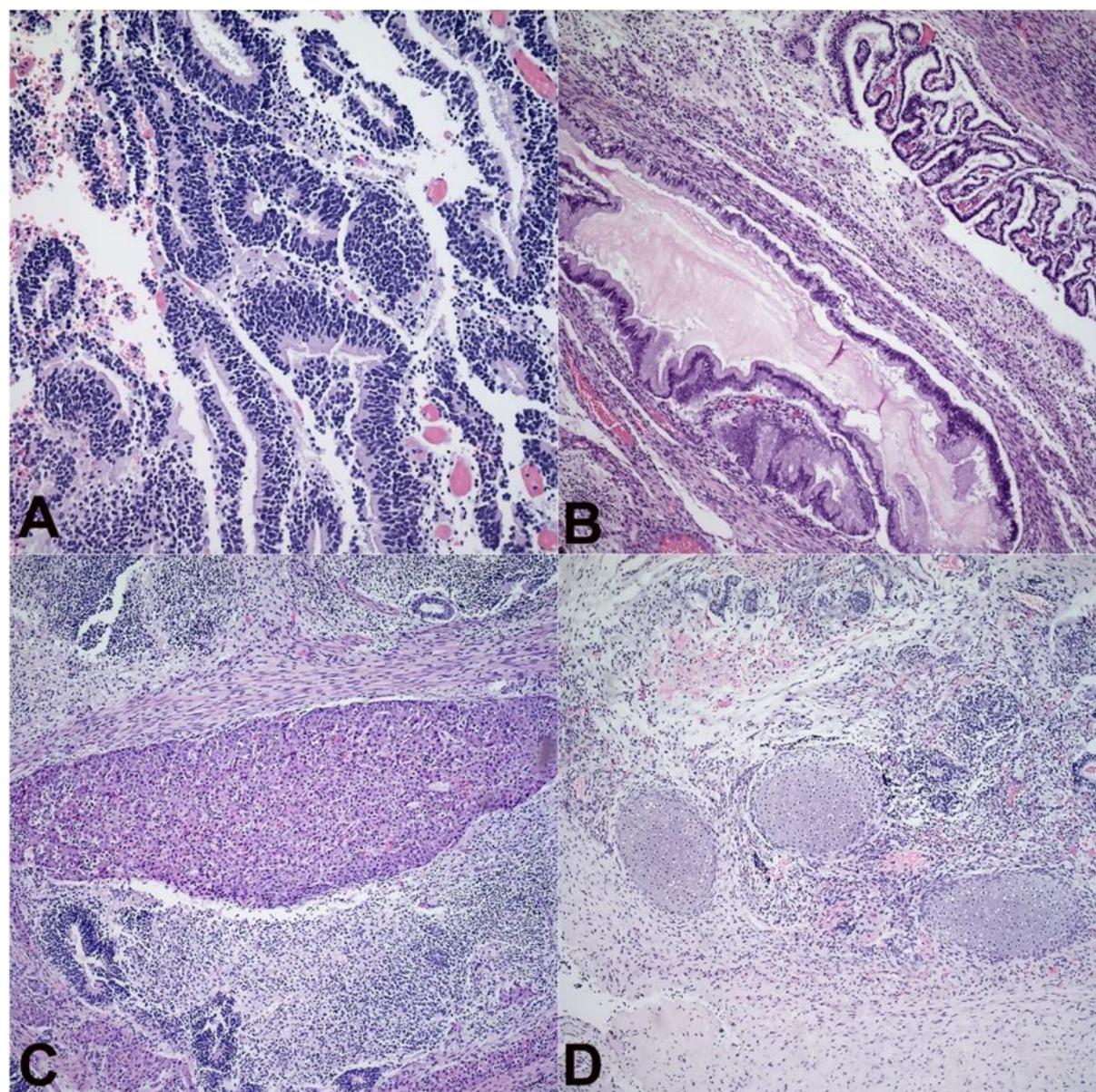


Figure 6. Immature neural tissue (**A** – H&E, 200X), Epithelial component (**B** – H&E, 100X), Adrenal tissue (**C** – H&E, 100X) and mesenchymal component (cartilage) (**D** – H&E, 100X) identified in representative sections of the mass.

Table 1. Organ weights and measurements of the patient compared to standards⁸

	B Weight (g)	Thymus (g)	Heart (g)	Lungs (g)	Spleen (g)	Liver (g)	Kidneys (g)	Adrenals (g)	CR (cm)	CH (cm)	HT (cm)
31 weeks	1375±281	4±3.4	9±2.8	28.5±13.2	4±1.2	55.4±17.3	13.7±5.2	3.7±1.3	27.6±3.8	37.8±3.1	5.9±0.7
32 weeks	1543±519	4.7±3.6	10.1±4.4	30.2±19	4.7±5.4	62.5±30	15.2±7.4	4.1±1.7	28.4±9.5	38.9±5.7	6.1±1.1
Patient	2494	84.15	9.95	45.03	5.75	89	17.54	4.16	31.5	43.9	6

B= body; CH= crown heel; CR= crown rump; HT= heel toe.

DISCUSSION

Among pathologic mediastinal tumors of infants and children, germ cell tumors (GCTs) are third most frequent after neurogenic tumors and foregut cysts.⁹ Neurogenic tumors most commonly arise in the posterior mediastinum, while the most frequent GCT of the anterior mediastinum in neonates and infants are teratomas.¹⁰ Teratomas arise from the aberrant differentiation of fetal germ cells. Normal migration of these abnormal germ cells causes gonadal teratomas, whereas abnormal migration leads to extragonadal tumors.¹¹ In the pediatric age group, mediastinum is the second most common site for extragonadal germ cell tumors (GCTs), with the sacrococcygeal region being the most common.⁵

The space occupying properties of thymic teratomas are responsible for its deleterious effects. Respiratory distress is the most prevalent symptom. In utero, the fetus may develop pericardial effusion, cardiac tamponade, and non-immune hydrops fetalis. Our patient was hydropic, hypotensive, and in respiratory distress. He had severe hydrops manifesting as anasarca.

The underlying mechanism of hydrops in thymic teratoma is impaired venous return. Also, airway obstruction leads to dilation of pulmonary veins and resultant impaired cardiac filling. Autopsy findings in our patient clearly depicted these mechanisms. Highly vascular teratomas can cause hydrops by additional mechanism of arteriovenous shunting and/or massive hemorrhage. This mechanism is, however, more common in sacrococcygeal teratomas.

Thymic teratomas may be discovered incidentally. CT nonetheless is the mainstay of diagnosis. Appearances on chest radiography are usually indistinguishable from many of the other causes of anterior mediastinal masses including bronchogenic cysts, lymphomas, lymphangiomas and neuroblastomas. Histological examination is therefore necessary for diagnosis. In our patient, the chest radiograph showed an expanded cardiothymic silhouette which was worrisome for a mediastinal mass, however, a CT or MRI could not be performed due to patient's clinical instability.

Thymic teratomas resemble gonadal teratomas histologically as well as immunohistochemically. Grossly they may be solid or cystic. Cystic areas

usually contain sebaceous material, hair, and teeth. Histologically, teratomas are composed of multiple cell types derived from more than one germ cell layers. They can be mature or immature. Immature teratomas usually contain immature neuroepithelial tissue such as immature brain tissue or embryonic tubules. They are graded based on degree of immaturity, presence of a neuroepithelial component and the quantity of the latter.¹² Our patient's teratoma contained abundant immature neural elements and was therefore grade 3.

Delayed or missed diagnosis can have serious implications including morbidity and mortality. Prenatal imaging aids early diagnosis. Surgical excision is usually curative and can even be performed in-utero.¹³ Our patient received aggressive medical management, but unfortunately an early surgical intervention could not be performed. Most congenital/neonatal teratomas are histologically benign with low potential for malignant transformation.¹⁴ The presence of immature elements, although worrisome, has no bearing on prognosis.¹² NIHF and pulmonary hypoplasia, however, are the primary factors contributing to fetal or neonatal morbidity and mortality.¹³ In our patient, in addition to NIHF and hypoplastic left lung with early signs of hyaline membrane disease, functional abnormalities of the heart, unabating anemia, leukopenia, and acidosis culminated in a poor outcome.

CONCLUSION

Thymic teratoma is not an infrequent cause of NIHF. It should be high on the list of differential diagnoses in a neonate with NIHF and expanded cardiothymic silhouette on chest radiograph. Early surgical intervention can be life-saving in these cases.

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