Superior Herniation of Normal Mediastinal Thymus

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Abstract

Superior herniation of normal mediastinal thymus is a rare cause of anterior neck swelling and only few cases are reported in literature. It clinically presents as a suprasternal swelling in the neck that appears during increased intrathoracic pressure. It is a benign condition that requires clinical and radiological awareness to avoid unnecessary investigations and invasive procedures. Ultrasound is the imaging of choice for the diagnosis and the management is conservative. This is a case report of a 3-weeks-old male neonate who presented with anterior neck swelling and stridor, diagnosed as a case of superior herniation of normal thymus and was managed conservatively.

Keywords: superior mediastinum, thymus, herniation, anterior neck swelling, ultrasound, increased intrathoracic pressure, Valsalva maneuver.

Introduction

Superior herniation of normal mediastinal thymus is an intermittent migration of the broadest part of normal mediastinal thymus into the suprasternal region during increased intrathoracic pressure.[1,2,3,4] It is a rare clinical entity and a least considered cause of anterior neck mass due to lack of awareness, leading to extensive unnecessary investigations and interventions. Thymus is necessary for normal immune function in the pediatric age group. It is important to differentiate superior herniation of normal thymus from other causes of anterior neck swelling particularly ectopic cervical thymus, to avoid unnecessary surgical excision. Therefore, high level of clinical suspicion and awareness are of paramount importance.[5]

Case Report

A full-term male neonate with insignificant birth history, who remained clinically well and discharged on the second day of life, presented at 3 weeks of age to Emergency Department of Sultan Qaboos University Hospital (SQUH) with history of anterior neck swelling that was only visible during crying and was associated with noisy breathing since the age of 2 weeks. There was no history of apnea, cyanosis, feeding difficulties, or respiratory distress. On examination, there was an anterior midline neck swelling arising from the suprasternal notch and extending up to the mid of the neck, visible only during crying and associated with stridor. (Figure 1). The swelling was soft in consistency and non-pulsatile. There was no bruit, and overlying skin appeared normal. Other systemic examination was unremarkable except for bilateral hydrocele. There was no dysmorphism, and he was thriving well with anthropometric measurements at 50th percentile for age and sex. Our differential diagnosis included apical lung herniation, ectopic cervical thymus, and laryngocele. Chest X-ray revealed the apical part of right upper lobe was hyperinflated. Computed Tomography (CT) angiogram of the neck and thorax ruled out congenital and vascular anomalies; there was a bulky thymus, normal for his age. Laryngobronchoscopy revealed normal upper and lower airway anatomy. A noticeable

bulge was observed at suprasternal region which appeared during inspiration and disappeared during expiration, raising the possibility of pleural bulging or apical lung herniation; however, on clinical examination, there was no correlation between the swelling and normal breathing. Fluoroscopy showed an intermittent appearance of soft tissue lesion in the suprasternal region anterior to the trachea. (Figure 2). Finally, an ultrasound imaging of the neck was done which revealed a soft tissue lesion arising from the superior mediastinum, solid in nature and in continuity with the thymus (Figure 3)**.** The diagnosis of superior herniation of normal thymus was established. The patient was managed conservatively and discharged in a stable condition. On follow up at the age of 14 months, he remained well and thriving with no signs of respiratory distress or stridor noted. The mother reported on phone follow-up that by the age of 4 years, the swelling was no longer obvious during straining or Valsalva maneuver. (Figure 4).

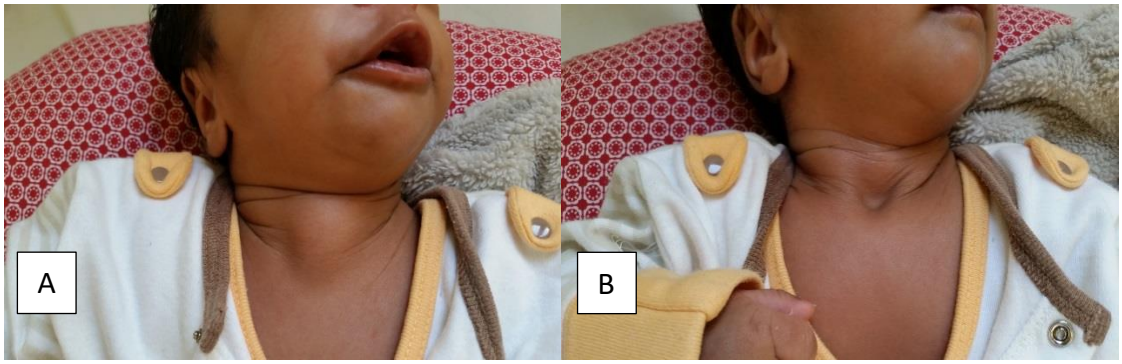

Fig. 1- (**Fig. 1A):** Appearance of suprasternal swelling during crying (Superior herniation of normal thymus during raised intrathoracic pressure). (**Fig. 1B**): Disappearance of swelling when the child is quiet

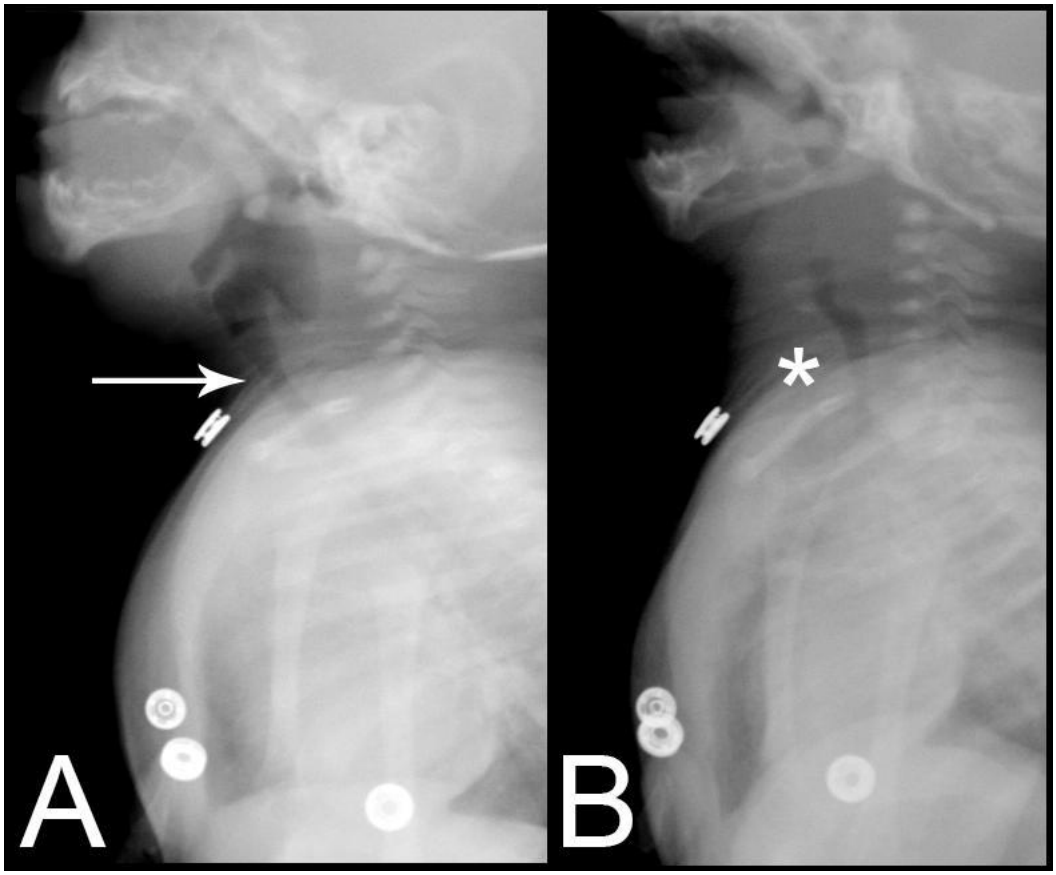

Fig. 2: Fluoroscopic examination of the neck in lateral view reveals an intermittent appearance of a soft tissue lesion (asterisk) in the suprasternal region- anterior to the trachea **(Fig. 2B)** displacing the trachea posteriorly. The normal appearance of the trachea (arrow in **Fig. 1A**) without the presence of suprasternal soft tissue lesion is shown in (**Fig. 2A).**

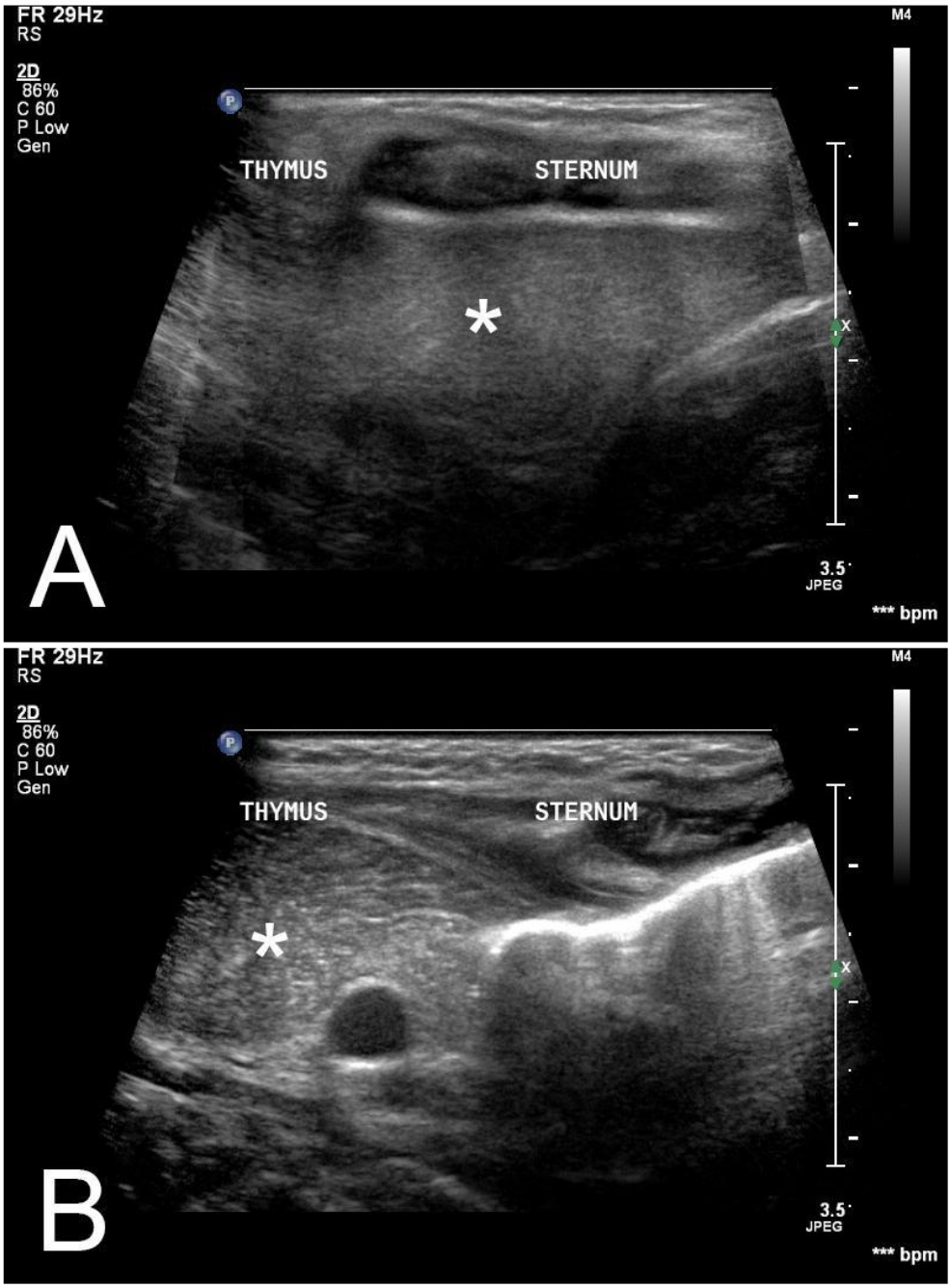

Fig. 3- (Fig. 3A): Midline sagittal ultrasound image using a high frequency linear transducer (5-12 MHz) through the sternal window reveal a homogeneous hypoechoic structure (asterisk) in the retrosternal region with internal echogenic foci "giving starry sky appearance", a normal sonographic morphology of the thymus in a neonatal period. (**Fig. 3B)** shows interval intermittent displacement of the thymus in the suprasternal region in the midline anterior to the trachea. No other soft tissue mass was visualized. The findings are consistent with intermittent suprasternal herniation of the normal thymus gland.

Fig. 4. (Fig. 4 A): No obvious suprasternal swelling at rest. (**Fig. 4 B):** No obvious suprasternal swelling during straining.

Discussion

This 3-week-old male neonate presented with anterior neck swelling at the suprasternal region apparent only during crying and associated with stridor was diagnosed with superior herniation of normal thymus.

Superior herniation of normal thymus is a benign cause of anterior midline neck swelling and requires no surgical intervention as thymus involutes with age. [2] It is defined as an intermittent herniation of the broadest part of normal thymus in suprasternal region out of superior mediastinum during raised intrathoracic pressure.[1,2,3,4] This abnormal movement of the thymus is possible because of loose connective tissue surrounding the thymus.[3,6] It is a rare clinical entity and only few cases are reported in literature; however, its incidence on ultrasonography is reported as 9%. [6] There is no particular gender predilection, however, more common in younger children as reported by Norma et al..[7] Familial pattern has been reported by Senel et al..[1] Other causes of neck masses that appears only during increase intrathoracic pressure

include apical lung herniation, jugular phlebectasia, and laryngocele. All of these are easily differentiated from each other through imaging.[2]

It is crucial to differentiate benign superior herniation of the normal mediastinal thymus from ectopic cervical thymus, which is due to migrational defect during thymus embryogenesis and has a tendency for malignant transformation. Surgical removal and histological examination are required for the establishment of diagnosis of ectopic cervical thymus.[5,8] Characteristic history, examination and ultrasonography led to definitive diagnosis of superior herniation of normal thymus and avoidance of unnecessary invasive investigations such as biopsy and surgical removal. [3] In classical cases, the parents primarily report's a noticeable swelling in the neck during crying or straining of their child that reproducible with Valsalva maneuver on examination in older children. [3] Ultrasonography as the first imaging of choice reveals a midline suprasternal soft tissue with echogenicity and echo pattern similar to that of normal thymus and seen in direct continuity with the thymus in upper mediastinum. [6,9,] However, in cervical ectopic thymus anatomic continuity between normal thymus and ectopic tissue could not be demonstrated and Tru-cut biopsy is required to confirm the diagnosis. [3,4] MRI is helpful to evaluate for any compression on adjacent structures.[3]

This neonate had an anterior midline neck swelling in suprasternal region which appeared during crying and disappeared at rest and during sleep. It was also associated with stridor, so laryngobronchoscopy and CT angiogram were done to rule out other structural abnormalities of airway. We diagnosed using ultrasonography and managed him conservatively. Badawi et al. reported a case of superior herniation of thymus in a preterm female infant who was born at 26-week gestation age and presented at 38-week corrected gestation age with similar clinical picture and diagnosed on ultrasound. [10] McDougall et al. reported an 8-month- old male infant

who presented with 3-month history of intermittent anterior midline neck swelling only during straining and was diagnosed by airway fluoroscopy rather than ultrasonography.[6]

No surgical intervention is needed for superior herniation of normal mediastinal thymus, and it is expected to disappear with time as depicted in our case. [6,10]

Conclusion:

Herniation of normal thymus is a least considered cause of anterior neck swelling among physicians and radiologist as it is very rare. However, with the awareness of its clinical presentation, normal anatomy and radiological findings, we can avoid unnecessary extensive investigations, misinterpretation and even surgical excision. The authors recommend that, for any child with anterior midline suprasternal swelling that appears during increased intrathoracic pressure and disappear at rest, radiological investigation with X-ray followed by ultrasound must be carried out before opting for more advanced investigations and invasive procedures.

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