□ Case Report □

# A case of true thymic hyperplasia in the mediastinum with ectopic thymus in the neck

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True thymic hyperplasia and ectopic thymus are very rare in children. In embryologic aspect, thymus is distributed around cervical area and ends up in mediastinum. This case is simultaneous thymic hyperplasia of neck and mediastinum. Ectopic thymus in the neck and thymic hyperplasia in the mediastinum in children were reported 2 and 7 cases respectively in Korea. In Clinical aspects, these thymic hyperplasia were presented by mass. So we should suspect these benign condition to avoid unnecessary operation or biopsy. We report a case of true thymic hyperplasia in the mediastinum with ectopic thymus in the neck in a 4-month-old male infant and review the relevant literature. We believe this is the first reported case in the world of true thymus hyperplasia in the mediastinum with cervical ectopic thymus in the neck. **(Korean J Pediatr 2006;49:996-999)**

**Key Words :** True thymic hyperplasia, Ectopic thymus

## Introduction

The thymus, which is at its largest during puberty, weighs approximately 30 g. It is composed of lymphocytes and decreases gradually in size after puberty[1, 2].

Cardiomegaly is not easily distinguished from enlargement of the thymus or from a tumor in the anterior mediastinum[3]. Thymic hyperplasia may be classified into two categories[1, 4-6]. True thymic hyperplasia is characterized by normal thymus structure, but the thymus can weigh more than 10% of the subject body weight. Thymus enlargement is not induced by other sources of systemic stress. Such true thymic hyperplasia is very rare in the world[1]. In the other type of thymus hyperplasia, known as lymphoid or follicular hyperplasia, which is frequently related to myasthenia gravis, the thymus is only slightly larger than the normal thymus. Lymphoid or follicular hyperplasia is characterized histologically by the presence of an activated germinal center of lymphoid follicles.

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In addition, ectopic thymus detected in the neck is very rare in children and requires diagnostic differentiation from neck tumor and other diseases via a sonogram. In Korea, 2 cases of ectopic thymus in the neck were reported until now[7, 8]. In Korea, 7 cases of thymic hyperplasia in the mediastinum were reported until now in the children[9-11]. Here, we report a case of true thymic hyperplasia in the mediastinum with ectopic thymus in the neck in a 4-month-old male infant and review the relevant literature.

## Case report

A 4-month-old male was admitted in Gyeongsang National University Hospital with a mass in his neck. The patient had irritation in the neck and rhinorrhea, which began 2 weeks prior to admission; he also received treatment for diarrhea 1 week prior to admission. During treatment for an upper respiratory infection, a 5.4 cm-sized, hard mass without rebound tenderness in the left submandibular location was detected accidentally, and thus he was transferred to Gyeongsang National University Hospital.

His past medical history was as follows. Because of upper respiratory infection, he underwent a simple chest x-

ray 1 month ago. At that time, cardiomegaly was not detected. Family history was nothing noteworthy. At the time of admission, the child appeared to be relatively healthy, with a blood pressure of 90/41 mmHg, a pulse rate of 120 beats per min, a respiratory rate of 32/min, and a temperature of 37.2℃. His weight was 7 kg (25 percentile) and hight was 65 cm (25-50 percentile). In the right submandibular location, a hard neck mass of approximately 3.3 cm without rebound tenderness was detected; other special features were not detected. Peripheral blood test results were as follows: hemoglobin was 11.7 g/dL, red blood cell volume was 35%, white blood cell number was 14,220/$\mu$L (polymorphonuclear cells 30%, lymphocytes 57%, mononuclear cells 7%), platelets were $197\times10^3/\mu$L, red blood cell sedimentation rate was 20 mm/Hr, PT/aPTT was 11.3/36.4 sec, and c-reactive protein was 12 mg/L. Blood biochemical results were AST : 46 U/L and ALT : 25 U/L. Other than the neck mass shown by MRI (Fig. 1) and a mass in the chest shown by CT pictures (Fig. 2), special features were not detected. The MRI of the neck detected an approximate 6.7 cm tumor with an intensity identical to that of the liver. The chest MRI detected a mass from the anterior mediastinum to the base of the heart, but primarily in the anterior mediastinum, with an intensity identical to that of the liver. Contrast enhancement was not detected. The mass in the chest and the neck mass were not connected (Fig. 3). The neck mass was 5.1×2.3×4 cm in size and 26 g in weight. The right thymus (to the right of the mediastinum) was approximately 7.5×6.2×3 cm in size and 46 g in weight and the left thymus (to the left of the mediastinum) was approximately 8.8×6.5×1.2 cm in size and 47 g in weight. Microscopic findings were identical to normal thymus tissue. The neck mass was an ectopic thymus and the chest mass was a true thymic hyperplasia.

Because thymoma and ectopic thymus were suspected, the child underwent a neck sonogram and magnetic resonance imaging. On the 8th day of admission, a fine needle aspiration biopsy was performed with sonogram monitoring, but an adequate amount of tissue was not obtained. Again, on the 22nd day of admission, fine needle aspiration biopsy was performed with monitoring by a sonogram in order to obtain a differential diagnosis and confirmation of

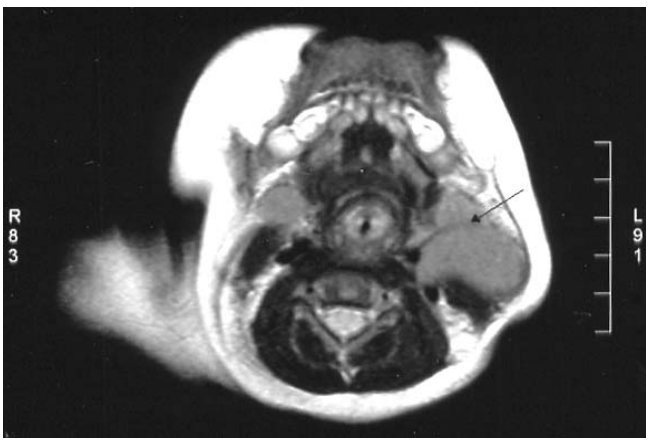


**Fig. 1.** Neck MRI reveals a 5.1×2.3×4 cm sized, benign looking soft tissue mass on the left side of the neck.

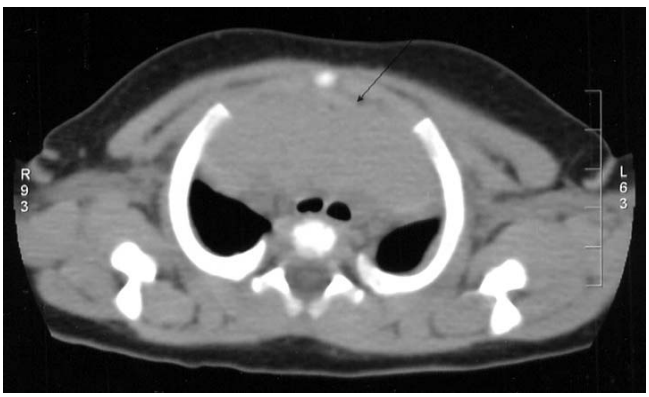


**Fig. 2.** Chest CT reveals a huge mediastinal mass.

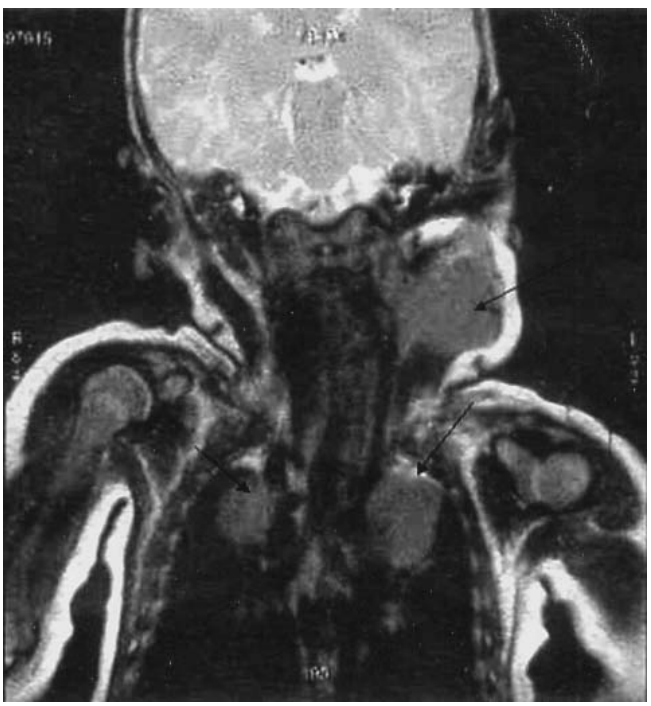

**Fig. 3.** Neck and chest MRIs reveal that true thymus hyperplasia and ectopic thymus were not connected.

lymphoma and thymoma. Open thoracic surgery and excisional biopsy of the neck mass were performed. The patient was subsequently discharged with a good prognosis.

## Discussion

Platter described thymic hyperplasia in infants as the enlargement of the thymus, which compresses the airway, resulting in mors thymica or "sudden infant death"[5]. The diagnosis of thymic hyperplasia is based on the weight and size of the thymus : the thymus is bigger than the predicted maximum range for the age[6] or the thymus weighs over 100 g[12]. According to a 1994 report, cellular growth is similar in thymic hyperplasia and the normal thymus; thus, thymic hyperplasia is a benign enlargement of the thymus[13]. Tumors that develop in the anterior mediastinum must be differentiated. The first type, thymomas are a characteristically malignant type of thymic hyperplasia that induce symptoms of pain or superior vena-cava syndrome and medullar differentiation of the parenchyma similar to malignant tissues formed in the cortex. The second type, proliferation of lymphoid follicles, is characterized by a thymus of normal size composed of lymphoid follicles with histologically activated germinal centers. In contrast to true thymic hyperplasia, this condition is often related to myasthenia gravis. The third thymolipoma type is characterized by the presence of binary lobes detectable by computerized tomography, and mature adipose tissue is present histologically. The fourth type, lymphoid lymphoma, is the most frequent tumor in the mediastinum in children and is difficult to diagnose with the small amounts of tissue that are usually obtainable by fine needle biopsy or fine needle aspiration biopsy; differentiation is necessary in order to start anti-neoplastic treatment for lymphoma[5, 13, 14]. Thus, to differentiate lymphoid lymphoma from other tumors in the thymus or from tumors that develop in the anterior mediastinum, dissection for histological diagnosis may be unavoidable[1]. Ectopic thymus is an extremely rare disease, but has been reported by several investigators recently. In fact, use of sonography may be responsible for its increased diagnosis in children[15]. With respect to the pathophysiology of the disease, if the embryonal thymus remains in the cervical areas, rather than descending from the cervical areas, the thymus may become a mass or even undergo malignant transformation[16]. Open thoracic surgery and dissection of the mass in the mediastinum were performed to differentiate thymoma from lymphoma in our patient. A frozen biopsy, followed by total dissection, was performed, and the patient has remained healthy since then. Selective hypogammaglobulinemia has developed in some cases following surgical removal, although the condition has not been associated with adverse health effects for 2 years in the children.

In embryologic aspect, thymus is distributed around cervical area and ends up in mediastinum. Clinically, it could be positioned at cervical area, so its remnant might induce hyperplasia, which is even rarely found as ectopic thymus at cervical area. And thymus that arrives at mediastinum may cause hyperplasia, which is even rarely diagnosed as mediastinal thymic hyperplasia on clinical background. But our patients showed hyperplasia at these two areas simutaneously. This case is a exceptionally rare phenomenon as unprecedented in clinical reports. Thus, we could not compare cases under examination. However, clinical identification of each disease will need corresponding skills and a further understanding about pathophysiology that causes this case. Ultimately, a case with true thymic hyperplasia that cannot be distinguished from other diseases should be operated. Lately, the policy for thymic mass in the mediastinum is that we did not perform unnecessary operations by using steroid on a short terms. It is estimated that our infant cases could be also recovered more even without any operation by using short-term steroid. But we found it necessary to check up on their pathology, since there was not any case of hyperplasia in two areas simultaneously at that time.

We believe this is the first reported case in the world of true thymus hyperplasia in the mediastinum with cervical ectopic thymus in the neck.

## 한 글 요 약

### 종격동 진성 흉선 증식증에 동반된 경부 이소 흉선 1례

경상대학교 의과대학 소아과학교실

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**임재영 · 박찬후 · 우향옥 · 윤희상**

소아에서 종격동 진성 흉선 증식증과 경부 이소 흉선은 매우 드문 질환이다. 이에 저자들은 경부 종괴를 주소로 내원한 환아에서 조직학적 검사를 통해 종격동 진성 흉선 증식증에 동반된 경부 이소 흉선을 진단하였기에 문헌 고찰과 함께 보고하고자 한다.

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