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□ Case Report □

Intrathyroidal branchial cleft-like cyst in neonate

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A rare case is described of an intrathyroidal branchial cleft-like cyst in neonate. The patient was a newborn girl with a mass in the left lateral neck. The ultrasonography and computed tomography revealed a cystic lesion in the left thyroid. The lesion was enucleated surgically from the thyroid. Histologically, the cyst was lined by squamous or columnar epithelium and contained inflammatory cell infiltraion, thyroid and parathyroid tissue. The patient has been doing well without any evidence of thyroid dysfunction for 15 months. **(Korean J Pediatr 2006;49:1005-1009)**

Key Words : Branchial cleft-like cyst, Thyroid gland, Neonate

Introduction

Cysts and sinuses in the neck may be formed along the course of the first, second, third, or fourth branchial clefts as a result of improper closure during embryonic life[1]. Branchial cleft cysts are congenital epithelial abnormalities usually located in the lateral neck along the anterior portion of the sternocleidomastoid muscle. The cystic spaces are usually lined with squamous epithelium with keratinaceous debris filling the lumen, but respiratory-type epithelium can also be observed[2]. In infected or ruptured lesions, inflammatory cells are seen within the cyst cavity or the surrounding stroma. However, cysts with the histological features of branchial cleft cyst have been reported in unusual sites, such as oral cavity, parotid, thyroid, and pancreas[3-5]. Intrathyroidal branchial cleft-like cyst is extremly rare. Neonatal case of an intrathyroidal branchial cleft-like cyst, to our knowledge, has not been reported yet. This article studies the morphologic features of this unique occurrence in the thyroid.

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Case report

A 2,710 g female infant was born at 37 weeks of gestation, to a 26 year-old mother, who had no history of medication, radiation, or infection prenatally without history of abortion. The baby was born with normal vaginal spontaneous delivery as first baby, and admitted to our neonatal intensive care unit for further evaluation and management of a painless mass in the left anterolateral neck. The cyst of anterior neck was found in antenatal sonography at 25^{+1} weeks. The size was 2.2×1.9×1.3 cm. The cyst was uniloculated and hypoechoic in sonographic finding (Fig. 1). The APGAR score was 8 on 1 minute and 9 on 5 minute. The family history of hereditary or anomaly was not noted.

On admission, body temperature was 37℃, heart rate was 130 beats per minute, respiratory rate was 54 breaths per minute, and blood pressure was 60/45 mmHg without disparity between upper and lower extremities. The body weight was 2,710 g (50-75 percentile) and good activity and crying were observed. Anterior fontanel was open without bulging. The 3×3 cm sized mass was palpated at the left anterolateral neck under the left mandibular angle (Fig. 2). The mass was soft and well-demarcated. There was no heatness or tenderness. The breathing sound was clear and there was no chest wall retraction. The heart sound was regular and had no murmur. Abdominal wall

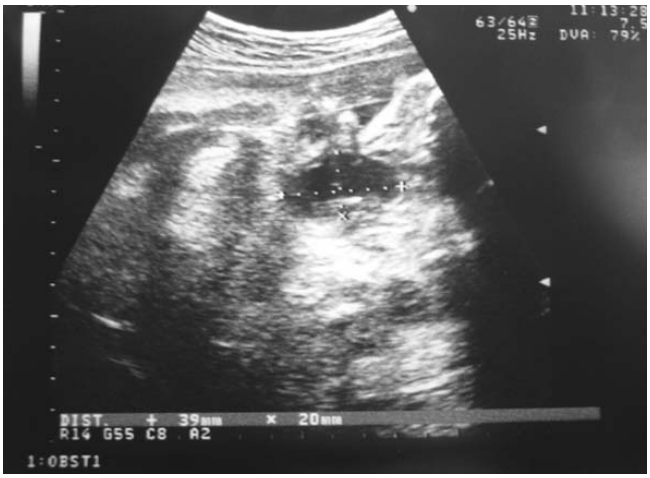

Fig. 1. 2.2×1.9×1.3 cm sized, anechoic and non-septated cyst on anterior neck. The movement at swallowing and inner flow were detected.

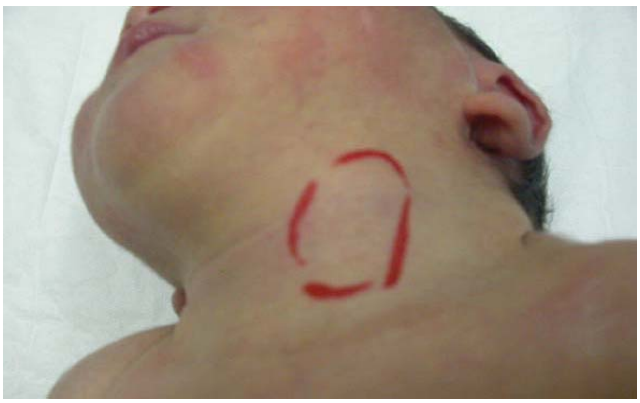

Fig. 2. 3×3 cm sized, soft and well-demarcated mass were palpated at Lt. anterolateral neck.

was soft. The bowel sound was decreased. Moro, sucking, and rooting reflexes were all good.

On admission, laboratory findings were as follows: 14,430/mm^3 of white blood cell, 15.4 g/dL of hemoglobin, 43.3% of hematocrit, 245,000/mm^3 of platelet, 30 IU/L of AST, 7 IU/L of ALT, 5.5 g/dL of total protein, 3.4 g/dL of albumin, 8.5/0.82 mg/dL of BUN/creatine, 76 mg/dL of glucose, 2.15 mg/dL of C-reactive protein and $Na^+/K^+/Cl^-$ 140/4.5/106.5 mEq/L. On gas analysis of arterial blood, the results were follows: 7.507 of pH, 31.5 mmHg of CO_2, 65.7 mmHg of O_2, 3.4 mmol/L of base excess, 96.4% of SaO_2, and 25.1 mmol/L of HCO_3^-.

On the chest X-ray, there was no cardiomegaly without increase of pulmonary vascular markings. On the ultrasonography, there were anechoic and well-demarcated cystic lesion in left thyroid gland, about 3.6×2.6×1.0 cm sized

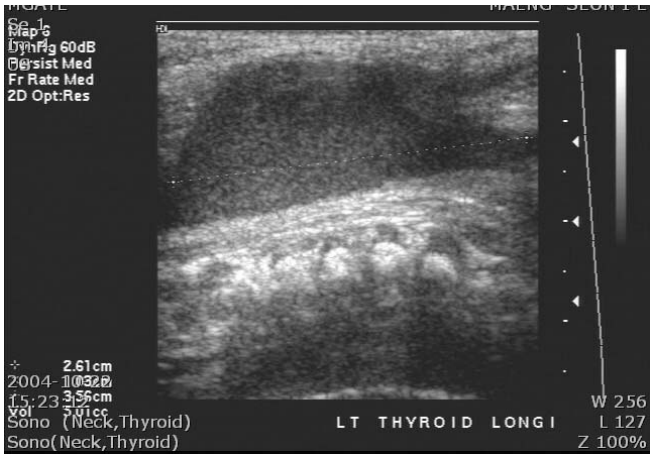

Fig. 3. On the neck sonography finding, anechoic and well-demarcated cystic lesion was noted at left thyroid gland.

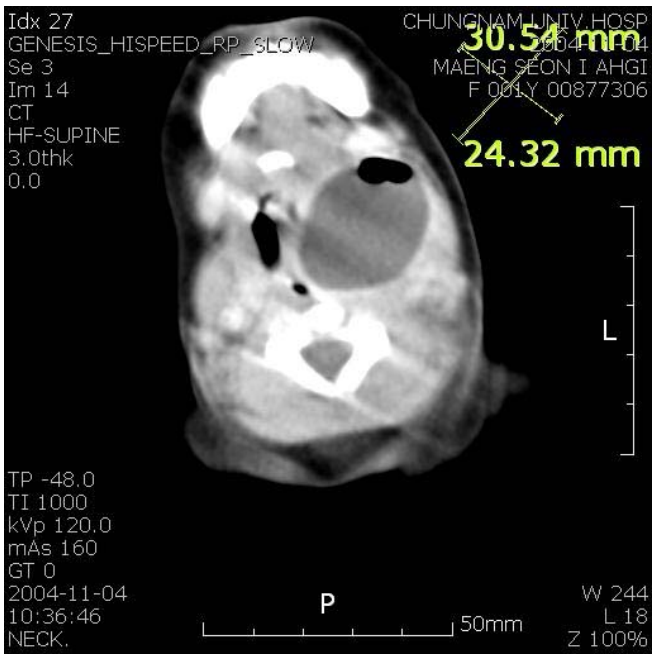

Fig. 4. On the CT finding, the cystic mass was containing high density materials, with contrast-enhancing even thickness peripheral wall in the left anterior neck.

(Fig. 3). On the computed tomographic scan, there was about 3×3×2.4 cm sized cystic mass, containing high density materials, with contrast-enhancing even thickeness peripheral wall in the left side anterior neck (Fig. 4).

Although the usage of the sulbactam sodium and ampicillin sodium and gentamicin sulfate from the second day of hospitalization, the size of mass was not decreased but increased on the fourth day of hospitalization, so we did aspiration examination of the mass. The inner content was yellowish and purulent. There was no squamous cell in

cytologic examination. The laboratory findings of the inner content were follows : 7.0 of pH, 180/mm^3 of red blood cell, 2,240/mm^3 of white blood cell (segment cell : 90%), 3.9 mg/dL of total protein, 8 IU/L of lactic dehydrogenase, 0 mg/dL of glucose, no growth of culture.

We did aspiration examinations several times, but there was no change in the size of the mass. The lesion was enucleated surgically from the thyroid on 17th day of hospitalization. Grossly, the specimen consists of a cystic mass measuring 3×3 cm in size. On section, it was uniloculated and the inner wall was irregular and colored dark red. Pathologically, a large cyst was lined with squamous or columnar epithelial cells, and a portion of thyroid and parathyroid tissue was also noted (Fig. 5). The cystic epithelial cells were dropped out from the wall by high graded inflammation (Fig. 6). After the operation, the Tc-99m Pertechnetate Thyroid Scintigraphy showed that the right thyroid lobe was enlarged and the left thyroid was not detected. The thyroid function tests were normal. She is 15 month-old and well without any evidence of thyroid dysfunction.

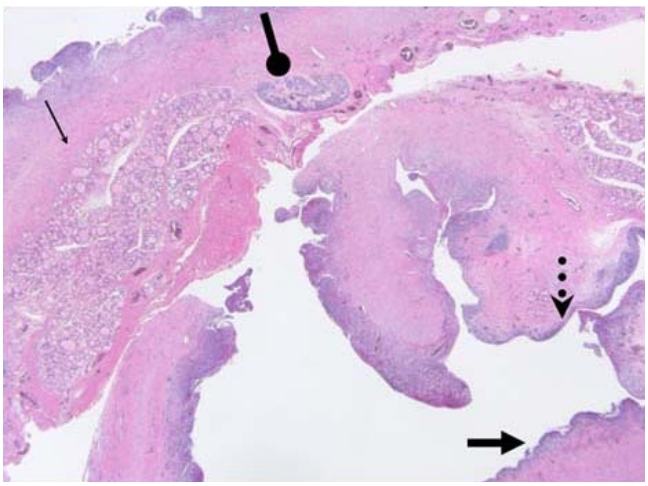

Fig. 5. A large cyst lined with squmous(dotted arrow) or columnar(arrow) epitherial cells. A portion of thyroid(narrow arrow) and parathyroid(rounded arrow) tissue is also noted (Hematoxylin & Eosin, ×12.5).

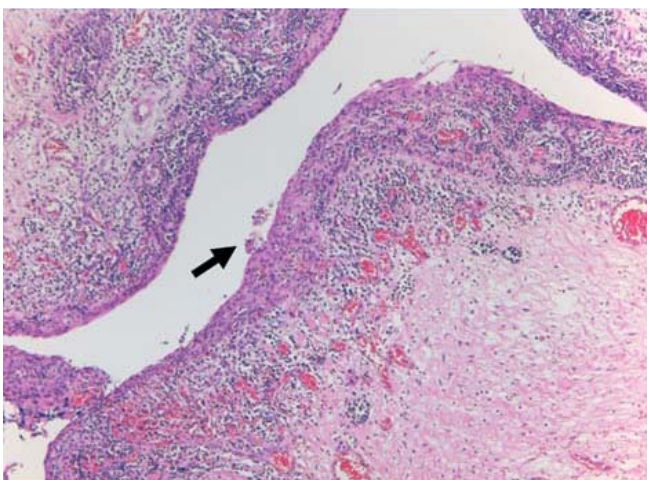

Fig. 6. A few epithelial cell nests(arrow) are detached from the lining because of severe inflammation(Hematoxylin & Eosin, ×12.5).

Discussion

A branchial cleft cyst is usually located in the lateral areas of the head and the neck and an intrathyroidal branchial cleft-like cyst is very rare. The term "branchial cleft-like cyst" has been used for an unusually located cervical lymphoepithelial cyst[3-9]. Only 15 cases, including our case, have been reported in the literature (Table 1)[6]. The patients ranged from 1 day to 71 years, with a mean age of 38.2 years. Except for our patient who was 1 day and another 7-year-old patient, all the other patients were adults. Among those patients were 12 female. It is very rare for an intrathyroidal branchial cleft-like cyst to occur in a male. Most of the cases occurred in women with a painless mass. The clinical symptom was usually a painless mass and four of the cysts were found incidentally. These lesions were usually unilateral and single. The multiple and bilateral cysts were rare and they have occurred in thyroids with papillary carcinoma. Thirteen (of 15) were associated with thyroidal disease, 4 with chronic lymphocytic thyroiditis, 3 with Hashimoto disease or chronic thyoiditis, 3 with nodular hyperplasia, 2 with chronic lymphocytic thyroiditis, nodular hyperplasia, and papillary carcinoma, and 1 with chronic lymphocytic thyroiditis and papillary carcinoma. Only two of the cysts, including the present case, occurred in the normal thyroid.

The cyst we described must be distinguished from other lesions in the neck, such as thyroglossal duct cyst, branchial cyst, and bronchogenic cyst. A thyroglossal duct cyst was excluded because of its location and histology despite no special differences in histological findings to branchial cleft cyst or thyroglossal duct cyst. It is usually located in the midline and usually connected with the hyoid bone, and it dose not have severe lymphoid cell infiltration with lymphoid follicles seen in branchial cleft cysts[8]. A branchial cyst was also excluded for its typical location in the lateral

Table 1. Reports of Intrathyroidal Branchial Cleft-like Cyst

Cases	Age/sex	Site	Size (cm)	Pathology (lining cells/associated conditions)	Symptom
Louis et al., 1989[8]	46/F	Bilateral lobe	0.6-1.2, 2.5	SC+focal CC, HT	Mass
Louis et al., 1989[8]	47/F	Left lower and upper poles	0.3-1.8	SC+focal CC, HT	Mass
Carney, 1989[11]	54/F	Right lower pole, multifocal	2.0	SC only, NH	Mass
Carney, 1989[11]	53/F	NA	0.5	SC only, NH	Hyperthy
Delabie et al., 1990[6]	34/F	Bilateral lobe	NA	SC only, NH	Mass
Radhi, 1994*	43/F	Left lobe	3.0	HT	Mass
Apel and Asa, 1994[3]	26/F	Left lobe	0.5	SC only, FVPC, CLT Heterotrophic thymus and parathyroid	Incidental
Apel and Asa, 1994[3]	39/M	Left upper pole	0.5	SC only, NH, PC, CLT	Incidental
Apel and Asa, 1994[3]	24/M	Left lower pole	0.7	SC only, CLT	Incidental
Apel and Asa, 1994[3]	52/F	Right lower lobe	0.3	SC only, CLT	Incidental
Apel and Asa, 1994[3]	58/F	Left lower pole	3×2.5×1	SC only NH, CLT, occult PC	Mass
Apel and Asa, 1994[3]	34/F	Right upper and lower pole	2.2, 1.5	SC only CLT	Mass
Haba et al., 2000[7]	71/M	Right upper pole	2.2×2.0×1	SC only, CLT	Mass
Park et al., 2004[10]	7/F	Left upper pole	2.9×1.8×4.2	SC+focal CC	Mass
Present case	1day/F	Left lobe	3×3×2.4	SC+focal CC	Mass

Abbreviations : HT, Hashimoto thyroiditis; NH, nodular hyperplasia; FVPC, follicular variant of papillary carcinoma; CLT, chronic lymphocytic thyroiditis; PC, papillary carcinoma; SC, squamous cells; CC, columnar cells; Hyperthy, hyperthyroidism.
*Original paper was not available; data from the article reported by Haba et al.[7]
Data from the article reported by Park et al., 2004[10]

neck, anterior to the sternocleidomastoid muscle[8]. The cutaneous bronchogenic cyst may be found in the neck even it has predominantly pulmonary or mediastinal location, especially in posterior to the carina[10]. The cyst of bronchogenic origin includes hyaline cartilage, respiratory epithelium, bronchial glands, smooth muscle, and nerve trunks in the cystic wall. However, this cyst was excluded in our case because of the intrathyroidal localization and the absence of smooth muscle and its connection with the hyoid bone.

The exact histogenesis of intrathyroidal branchial cleft-like cyst is unclear. It is probably derived from the development rests as thymic and parathyroid tissue is seen within the thyroid, and the relationship between the cystic enlargement of those rests and Hashimoto's disease[9]. In our case, the parathyroid tissue was observed, while the thymic tissue was not. There are two possibilities : either it is a developmental anomaly or an acquired lesion. The first possibility is that it might be a derivative of a developmental rest such as ultimobranchial body, solid cell nest, and branchial cleft remnant. The second hypothesis is a development from an acquired thyroidal diseases[3, 7-11]. Delabie et al. speculated that the branchial cleft-like cyst are derived from the solid nests related to the ultimobranchial body[7]. Solid cell nests have been found in up to 61% of adult thyroid[12], which were positive for carcinoembryonic antigen[13]. The lining of cyst consisted of squamous and columnar epithelium and contained thyroid and parathyroid tissue. Branchial cleft derivatives, such as thymus and parathyroid, develop in close association with the thyroid and may be found within the thyroid gland. These branchial cleft-like cysts also may arise from branchial cleft derivatives.

한 글 요 약

신생아에서 발견된 갑상샘의 아가미틈새양 낭종

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아가미틈새(branchial cleft) 낭종은 흔히 경부측면에 위치하나, 아가미틈새 낭종의 조직학적 소견을 보이는 낭종이 비전형적인 위치에서 발견되기도 한다. 갑상샘의 아가미틈새양(branchial cleft-like) 낭종은 이제까지 보고된 14례 중 7세 여아에서 발견된 1례를 제외하고는 모두 성인에서 발견되었다. 저자들은 좌측 경부의 종물을 주소로 내원한 신생아에서 초음파 검사, 컴퓨터 단층촬영, 병리조직학 검사를 통하여 진단된 갑상샘의 아가미틈새양 낭종 1례를 경험하였기에 보고하는 바이다.

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