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Case Report

A Rare Case of A Mass on the Neck that Changes its Dimensions When Weeping

Kamil Şahin[1], Murat Elevli[1], Fatma Saraç[2], Hatice Nilgün Selçuk Duru[1], Serdar Pop[1], Ayhan Koçak[3]

Abstract

When branchial clefts and pouches do not atrophy between 4 and 7 weeks of fetal life or when an anomalous or incomplete fusion occurs, congenital neck anomalies may develop. We present a rare case of a mass resembling a branchial sinus on the right side of the neck of a 2-month-old baby, which originated from the fourth branchial arch-derived structuresand varies in size with weeping. A 2-month-old boy was brought with a complaint of a congenital swelling on his neck. His physical examination revealed a cyst on the right lower region of the neck, which swelled when he wept, and prolonged neonatal jaundice. On computed tomographyof the neck, a formation consistent with a branchial cleft cyst originating from the right jugulodigastric region extending up to the right infraclavicular region and to the pyriform sinus was observed. The child was maintained under surveillance. Branchial cysts account for 30% of congenital neck cysts in children. Masses derived from the fourth branchial arch are more frequently seen on the left side. In our patient, this rare mass was localized on the right side of the neck and was diagnosed when the baby was 2 months old. During the Valsalva maneuver, pyriform sinuses fill with air passing through their orifices and consequently the size of the mass varies in size. Third and fourth branchial sinus anomalies should be considered in neck masses whose size changes with Valsalva maneuver.

Keywords: Branchial cyst, neck mass, valsalva

Introduction

Congenital neck anomalies are caused because branchial clefts and pockets do not disappear due to their abnormal growth and incomplete juncture between the 4th and 7th weeks in the fetus (1). Branchial cleft anomalies can be in the form of cysts, fistulas, and sinuses. While first and second cleft anomalies among the sinus structures usually open outward, the third and fourth sinus anomalies are associated with internal mucosal structures (2, 3). First cleft anomalies are usually seen in the form of fistulas and less often, in the form of cysts in the parotid gland (2, 3). Second branchial cleft anomalies are the most common among all branchial cleft anomalies and occur at a rate of 95%. They are usually cystic structures that can extend from the tonsillar fossa to the supraclavicular region in the submandibular triangle and enter the pharynx and tonsillar fossa. Third branchial cysts pass between the internal and the external carotid arteries in the neighborhood of the pharynx. Fourth branchial cysts are next to the pharynx, adjacent to the tonsillar pit, and near the carotid sheath (3, 4). The last two anomalies are very rare. We present the rare case of a branchial sinus mass originating from the fourth branchial structures and growing and shrinking in the right neck region due to crying in a two-month-old baby.

Case

A two-month-old male patient was admitted to the child polyclinic of Haseki Training and Research Hospital with congenital swelling in the right neck. He was Syrian and was the first child of a mother and father who were 18 and 20 years old, respectively; he was born at home. Through an Arabic translator, written informed consent was received from the father of the patient regarding the publication of the study. There was swelling in the lower right side of the neck, covering the entire neck region and extending to the clavicula, and its size changed on crying (Figure 1). The patient had extended newborn jaundice. He had natural breathing, a cardiac apex beat of 84/ rhythmic, and no murmur. His abdomen was comfortable, and no organomegaly was observed. The fontanel was 3×2 cm and natural. No features were observed in other systems. Externally, male testicles were detected in the scrotum. The patient had no difficulty breathing and feeding. He weighed 4200 g. During his full blood count and biochemistry examinations, he showed unremarkable features other than a total bilirubin level of 7.4 mg/dL. An irregularly contoured cystic lesion that was 4 cm in diameter was detected on performing superficial tissue ultrasonography of the right neck. No pathology was found on performing abdominal ultrasound. The branchial cleft cyst that started from the right jugulodigastric region and extended to the axillary level at the right infraclavicular level was lobule-contoured with a width of 45×32 mm at the widest area, did

[1]Department of Pediatrics, University of Health Sciences, Haseki Training and Research Hospital, İstanbul, Turkey
[2]Department of Pediatric Surgery, University of Health Sciences, Haseki Training and Research Hospital, İstanbul, Turkey
[3]Department of Radiology, University of Health Sciences, Haseki Training and Research Hospital, İstanbul, Turkey

Address for Correspondence:
Kamil Şahin
E-mail: drkamil_sahin@hotmail.com

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not have a pressure effect on vascular structures, was in the cystic form, and extended to the pyriform sinus (Fig. 2.a-c). By planning a detailed ear, nose, and throat examination and surgery under anesthesia, the patient was followed up by the pediatric surgery department.

Discussion

Among congenital neck cysts in children, 30% are branchial cysts. Their incidence has been reported to be 0.05%. While branchial cysts are equally seen among men and women and are more commonly among middle-aged people, fourth branchial cysts are seen during all ages and are more frequent in young babies. They can be accompanied by kidney, ear, and skeletal anomalies (5).

Between the intrauterine 4th and 7th weeks in embryogenesis, five pieces of ectodermal clefts and five pieces of the endodermal line among six pieces of the branchial arches composed of mesodermal tissues and the pharyngeal pouches that temporarily emerge are observed in the neck region. Congenital neck masses that belong to the first four of these six branchial structures can develop. The anomaly is located under the affected arch and is usually located in the region from the pre-auricular skin to the clavicula. Genetic susceptibility is thought to exist (4). The mandible, a part of the maxilla, inner ear, outer ear canal, and mastoid air cells are formed from the first branchial structures. The palatine tonsil, supratonsillar fossa, and a part of the hyoid bone are formed from the second branchial structures. The epiglottic arch, a part of the hyoid bone, paratroid gland, thymus, and aortic arch are formed from the third and fourth branchial structures. The fifth and sixth branchial structures are temporary, and no anomalies develop from them (1, 3, 4). Among the congenital branchial masses, 95% develop from the second branchial structures. Third and fourth branchial anomalies that are rarely seen can usually enter the pharynx through the pyriform sinus. The third cleft is on the superior laryngeal nerve. It extends from the supraclavicular fossa to the pyriform sinus. The fourth cleft is below the superior laryngeal nerve. It is mostly in the form of a sinus (6).

Figure 1. Branchial cyst in the neck

Although it is possible that they can be seen at all ages, the first and second branchial structures are seen in the middle-age group and the third and fourth branchial structures are seen during the first 10 years of life. In our patient, the mass that extended from the right supraclavicular fossa to the right pyriform sinus was also thought to originate from the fourth branchial structure. There was no history of a similar structure in the family. There were no additional ear, skeletal, or renal anomalies. Fourth cleft anomalies can pass down from the pyriform sinus to the mediastinum. Most third and fourth branchial cleft anomalies are located on the left side and spread to the pyriform sinus (7). An endoscopic examination under general anesthesia was not performed because the patient was young. Treatment for all cleft anomalies is surgical resection. Our patient was followed up by the pediatric surgery department, and the operation was planned.

Fourth branchial masses that are very rare are mostly located on the left side. Our case is a rare case because it was located on the right side, he was diagnosed at the age of two months, and the size of the mass changed during the Valsalva maneuver. During the Valsalva maneuver, the mass enlarged and shrunk by air infiltration from the sinus mouths opening to the pyriform sinus.

Conclusion

In children, congenital anomalies of branchial structures should be keep in mind when neck masses are present, and third and fourth branchial sinus anomalies should be considered for masses that grow and shrink during the Valsalva maneuver.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this study.

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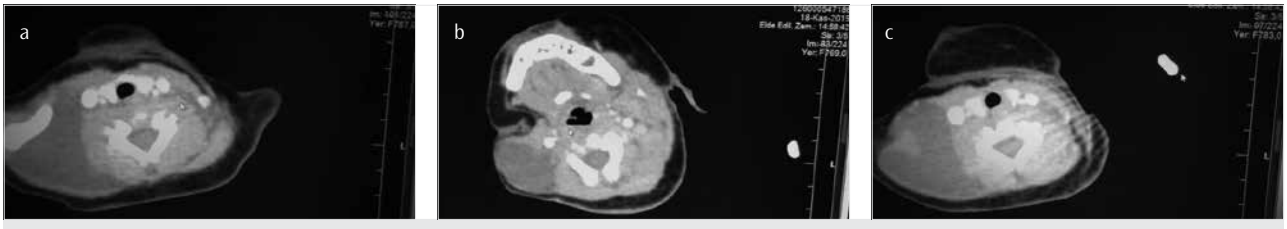

Figure 2. a-c. Hypodense cystic lesion which, starting from the right jugulodigastric region, extends from the posteromedial of the sternocleidomastoid muscle to the right supraclavicular fossa, and shows no contrast involvement

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