Ectopic salivary gland of the base of the tongue: a rare cause of neonatal respiratory distress

Mehmet İlhan Şahin[1], İbrahim Ketenci[1], Kerem Kökoğlu[1], Furkan Şan[1], İrfan Kara[1], Mehmet Adnan Öztürk[2], Olgun Kontaş[3]

Departments of [1]Otolaryngology and [3]Pathology and [2]Division of Neonatology, Department of Pediatrics, Erciyes University Faculty of Medicine. E-mail: dr.kokoglu@gmail.com

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Neonatal upper airway obstruction is a life-threatening condition and requires rapid assessment and effective treatment. Malformations of the upper airway of a newborn can lead to acute respiratory distress. The aim of the present paper is to report the case of a newborn with respiratory distress due to a tongue base mass, which was removed surgically. The patient needed a tracheostomy tube for only three days and then could breathe spontaneously. However, he had swallowing problems, which decreased gradually over 9 months. Histopathologically, the mass was found to consist of mucous salivary glands, and was recorded as an ectopic salivary gland, which is extremely rare among the types of masses that may cause upper airway obstruction in a newborn.

Key words: tongue, respiratory distress, newborn, ectopic tissue, salivary glands.

The region extending from the nasal aperture to the subglottic space is called the upper airway. Congenital anomalies of this region may lead to respiratory distress because of their size and location. Choanal atresia is the most common congenital cause of neonatal upper airway obstruction; bilateral vocal cord palsy is the second most common cause of stridor in neonates[1]. Along with these causes, pathologies of the tongue and tongue base, laryngomalacia, congenital subglottic stenosis, subglottic hemangioma and craniofacial anomalies such as Pierre Robin syndrome should be kept in mind as possible reasons for upper airway obstruction[1,2]. Congenital anomalies of the tongue are rarely seen in newborns. Anomalies of the tongue base such as hemangioma, lymphangioma, thyroglossal canal cysts, ectopic thyroid gland, ectopic salivary gland and hamartoma may be causes of respiratory distress[3]. Here we present the case of an infant born with respiratory distress because of a mass at the base of the tongue, who underwent surgical treatment.

Case Report

A 24-year-old mother gave birth to a boy by caesarean section at week 40 of gestation. She had no history of abortion. Polyhydroamnios was seen in the pregnancy, but no fetal anomaly was detected in the prenatal period. The newborn had respiratory distress and was intubated immediately. With intubation, he breathed spontaneously and did not need mechanical ventilation. There was no clue to any underlying syndrome. Upper airway examination revealed no nasal or oral pathologies, but there was an obstructive mass at the tongue base. The larynx and subglottic space appeared normal when direct laryngoscopy was performed (Fig. 1). The patient was able to breathe spontaneously and comfortably when the tongue base was elevated by the laryngoscope. When the tongue base was left to itself, stridor and supraclavicular and suprasternal retractions were seen. There was no cause of respiratory distress other than the mass in the tongue base; further examination was done for differential diagnosis. His serum level of thyroid-stimulating hormone (TSH) was 158.8 μU/ml (0.8-6.26), free thyroxine (T_4) 0.7 ng/dl (0.81-1.73) and free triiodothyronine (T_3) 2.08 pg/ml (2.76-4.94). The patient had hypothyrodism and was managed with

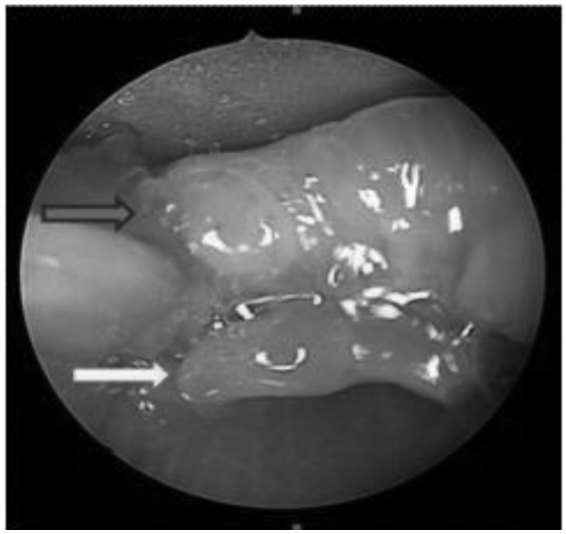


Fig. 1. A figure of tongue base mass during direct laryngoscopy, (Upper arrow is mass, lower arrow is epiglottis.)

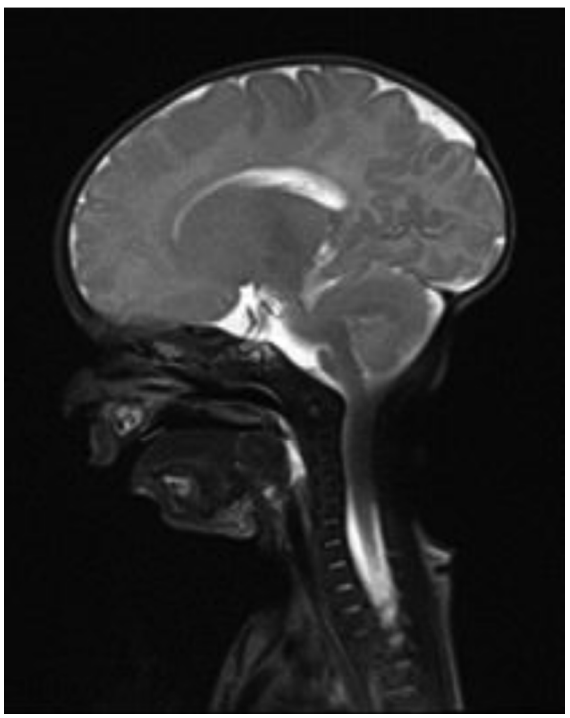


Fig. 2. MR imagination. There is no differenet density at tongue base.

levothyroxine; he still receives this treatment. The left lobe of the thyroid gland was atrophic, while the right lobe was intact and in its usual location in the ultrasound. No intensity different from that of the tongue was shown in the magnetic resonance imaging (MRI) (Fig. 2). In Tc99m scintigraphy, there was no

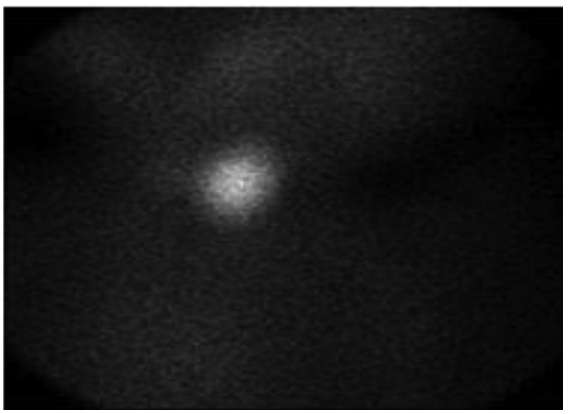


Fig. 3. Tc99-m scintigraphy.
Nuclear activation is only seen in right thyrold lobe

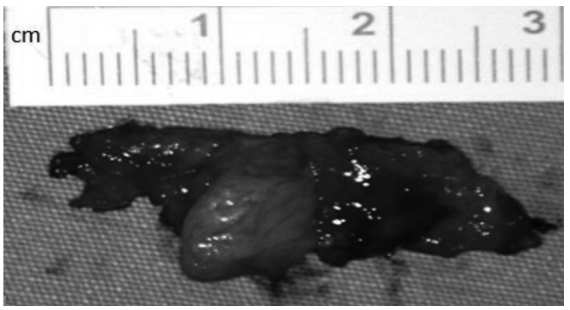


Fig. 4. A figure of specimen

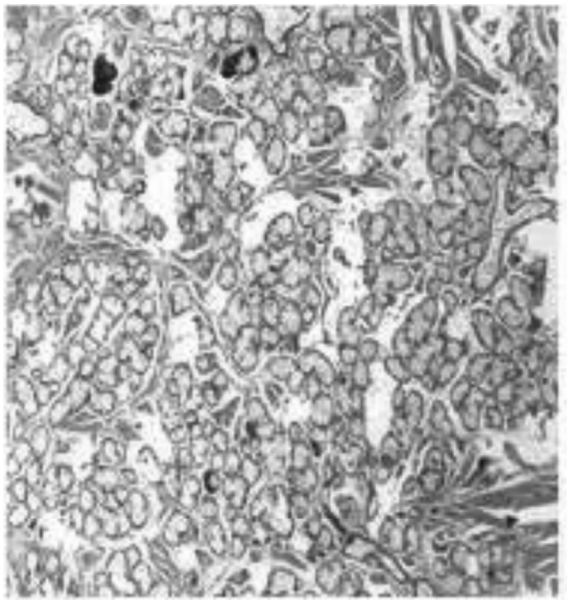
scintigraphic activity at the tongue base, and only the right lobe of the thyroid gland was active (Fig. 3).

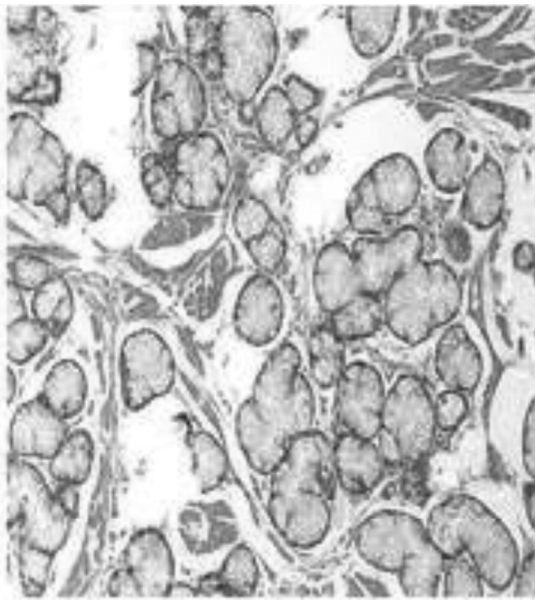
Surgery was planned for pathological diagnosis and treatment of the mass. Under general anesthesia, tracheotomy was done first to save the airway, and then the mass was excised endoscopically with a bipolar electrocautery. The specimen's size was 3 cm (Fig. 4). Upon histopathological examination, the specimen was found to be composed of mucous salivary glands, which had no signs of being atypical or composed of irregular tissue (Figs. 5,6).

The tracheostomy tube was removed on the 3rd day postoperatively. The spontaneous respiration was normal. There was no respiratory distress after decannulation; however, the patient suffered from dysphagia for weeks. Laryngoscopic examinations showed nothing but a slight fibrosis at the tongue base (Fig. 7).

Discussion

Congenital obstructive anomalies of the upper airway may be life threatening in newborns. Some of these anomalies can be related to





Figs. 5,6. Mucous salivary units are seen histopathologically. (Left: 40 times magnification. Right: 200 times magnification

the tongue. Pierre Robin syndrome is a major cause of obstruction due to the tongue. This syndrome was defined by Pierre Robin in 1923 as having three main features: glossoptosis, cleft palate and micrognathia[4]. Our case did not have malformations such as a cleft palate or micrognathia, so Pierre Robin syndrome was not considered to be the reason for the obstruction.

Whenever differential diagnosis is performed for masses of the tongue base, thyroglossal cysts and ectopic thyroid tissue should be kept in mind, especially in newborns. An ectopic thyroid gland is defined as one that is located in a position completely or partially different from its normal anatomical site. The most common ectopic thyroid gland location is the base of the tongue (90%)[5]. 33-62% of patients have hypothyroidism[6]. Radiologic methods such as ultrasound and Tc-99m scintigraphy are very helpful in the diagnosis of ectopic thyroid. The lack of a thyroid image in its normal anatomical region, and the presence of scintigraphic activity in the tongue base as detected by Tc-99m scintigraphy, are sufficient for clinical diagnosis. Our patient had hypothyroidism, but the right lobe of the thyroid gland was seen to be in its normal position on ultrasonography, and there was no scintigraphic activity at the tongue base. Thus, the diagnosis of ectopic thyroid was ruled out.

Thyroglossal duct cysts are the most common form of congenital neck cyst and are located at the midline of the neck[7]. These cysts commonly are found in the neck, and rarely in the tongue base. Fifty cases have been reported in the literature so far[8]. Computed tomography and MRI can be used for diagnosis. Radiologic studies show a smooth, well-defined homogeneously attenuating cystic lesion[8]. In our patient, MRI showed no lesion with a different density at the tongue base.

Carcinoma, hamartoma and ectopic tissue

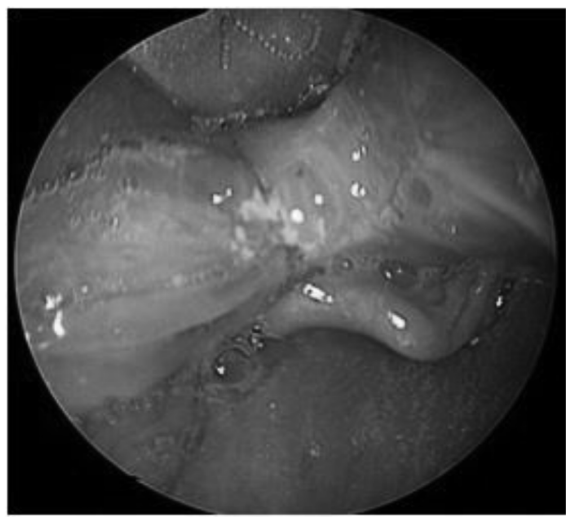


Fig. 7. A figure of postoperatively at first month. A fibrosis is shown at base of tongue

should also be considered in the differential diagnosis of masses of the tongue base[9]. Exact diagnosis is made histopathologically.

Tongue-base hamartomas are discussed in the literature[9,10]. A hamartoma is a benign, focal malformation that is composed of tissue elements normally found at the site in question, but which are present in a disorganized formation. The normal tissue of the tongue is formed by skeletal muscles and the stratified squamous epithelium that surrounds it. There are nerves, blood vessels, adipose tissue and minor salivary glands in the interstitial space[11]. The specimen from our patient was 3 cm in size; it showed mucous salivary glands formed typically, and no disorganized tissue. Minor salivary glands do exist at the tongue base[11]. However; a minor salivary gland cannot be 3 cm in size. For the reasons detailed here, the best diagnosis for this tissue was "ectopic salivary gland at the tongue base."

Ectopic salivary glands may be located in the hypophysis, middle ear, thyroid, parathyroid, lymph nodes or branchial cysts. Most of these lesions present early in life. The usual clinical expression is a draining sinus and swelling[12]. In our case, the ectopic gland caused respiratory distress because of its location and size.

Pathologies of the tongue base must be considered as a congenital cause of upper airway obstruction in newborns. Surgical procedures are successful in treating such masses. However, these patients will experience swallowing difficulties for a long time postoperatively.

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