Clinical case

Rare cause of respiratory distress syndrome in a child: Hamartoma of salivary gland in the nasopharynx.

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Abstract: *Relevance.* Hamartoma of salivary gland or salivary gland anlage tumor (SGAT) is an extremely rare pathology found mainly in children in the early months of life. *Description of the clinical observation.* This article presents a clinical case of respiratory distress syndrome in a 4-month-old child arisen due to a salivary gland hamartoma in the nasopharynx. The symptoms are nonspecific and are mainly represented by respiratory distress syndrome, difficulty in feeding, apnea, and sleep breathing disorders. SGAT is most commonly found in children in the first months of life, but there are cases of later onset. SGAT is a benign neoplasm, and surgical resection is the main treatment option. SGAT is characterized by the absence of recurrence after removal. The histological picture is a combination of epithelial and mesenchymal components. *Conclusion.* SGAT – a rare benign nasopharyngeal neoplasm in a child in the first months of life. Early diagnostics including endoscopic examination of the nasopharynx, CT and MRI examinations allowed us to determine the operative treatment tactics, achieve a good functional result in the form of complete relief of the child's complaints and prevent development of serious complications related to the upper airway obstruction.

Keywords: hamartoma of the salivary gland, respiratory distress syndrome, salivary gland anlage tumor, nasopharyngeal neoplasm in a child, congenital pleomorphic nasopharyngeal adenoma, benign nasopharyngeal neoplasm.

Introduction

Upper airway obstruction in infants is a frequent and potentially dangerous condition requiring hospitalization and careful examination. In most cases, these symptoms are non-specific and are present in the form of respiratory distress syndrome. The leading causes of persistent nasal airway obstruction in infants are choanal atresia and congenital deformities of the facial skull. Congenital neoplasms of the nasal cavity and nasopharynx are not frequent, but can also lead to marked nasal breathing disorders. A very rare congenital nasopharyngeal neoplasm presented in our clinical case is a hamartoma of salivary gland conception, or congenital pleomorphic adenoma found in a 4-month-old child.

Clinical case

In January 2023, patient N., 4 months 1 week old male, was admitted to the Children's Clinical Center of High Medical Technologies of St. Petersburg named after K.A. Raukhfus with complaints of shortness of breath, cough, and distant wheezing. It is known that he had previously been treated at the Research Institute of Pediatric Infections for acute bronchiolitis, but due to persisting symptoms of bronchoobstruction against the background of decreasing doses of inhalation drugs (Berodual, Budesonide) he was transferred to the specialized pulmonology department of the Raukhfus Children's Clinical Center of High Medical Technologies. According to his mother, in the first month of life the child had poor weight gain, noisy breathing through the nose. Due to her complaints, at the age of 3 months the child

underwent nasopharyngeal cavity and nasopharynx fibroscopy, but the report of nasopharyngeal neoplasm was not described, and the diagnosis was "deviation of nasal septum".

At the Children's Clinical Center of High Medical Technologies of St. Petersburg named after K.A. Raukhfus, a video tracheobronchoscopy was performed through a laryngeal mask, the conclusion being: bilateral catarrhal endobronchitis II stage with diffuse putreform hypersecretion and signs of impaired drainage function. CT scan of the thoracic cavity showed a picture of bronchiolitis.

Despite the treatment of the underlying disease received by the child, the complaints of nasal breathing difficulties persisted. In order to verify the cause of nasal obstruction, the child underwent a number of additional examinations. Under general anesthesia, the nasal cavity and nasopharynx were examined with a 2.7 mm fibroscope, as a result a volumetric smooth formation on a stem was found in the nasopharynx (Fig. 1 a, b), almost completely filling its lumen, balloting when breathing and prolapsing into the nasal cavity when exhaling.

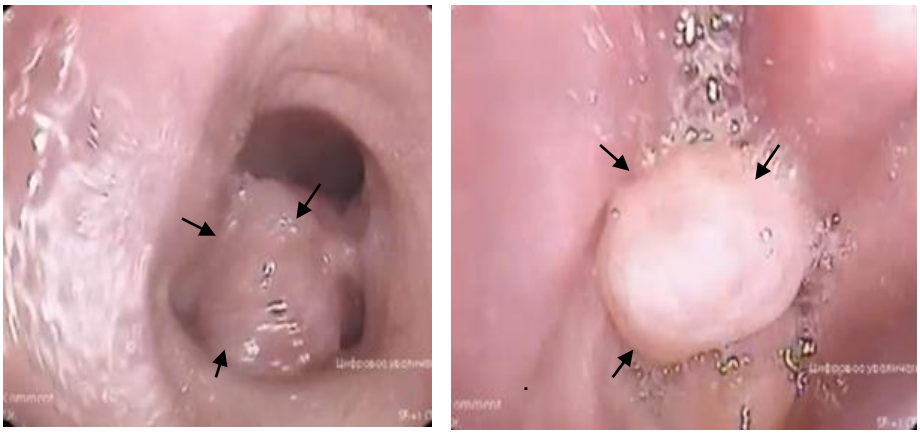

Figure 1. Image of the nasopharyngeal neoplasm (shown by arrows) during transnasal fibroscopy inhalation (a) and exhalation (b).

MRI of the brain with contrast revealed a volumetric mass in the anterior parts of the nasopharynx, rather homogeneous structure, isohypointensive MR signal on T1-, T2-VI, with indistinct, even contours, 17 x 12.5 x 15.5 mm in size. The mass is intimately adjacent to the soft palate, the pharyngeal tonsil, the border between the mass and the described structures being indistinct. The nasopharyngeal lumen at the level of the mass is not seen, the soft palate is pushed downwards causing narrowing of the oral cavity lumen.

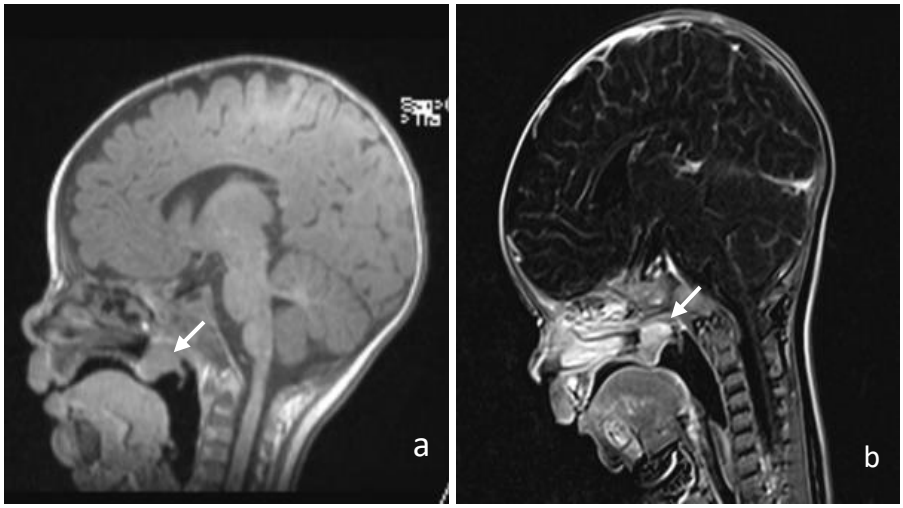

Figure 2. a) CT scan of the head and b) MRI of the brain with enhancement in the sagittal projection. The arrow shows nasopharyngeal neoplasm.

Based on the examinations performed, the child was consulted by an oncologist and the neoplasm was presumed to be benign. After stabilization of the underlying disease, the decision was made to remove the neoplasm in order to restore adequate nasal breathing. Under endotracheal anesthesia with endoscopic control (endoscopes 0° and 70°) after the preliminary soft catheter tying of the soft palate, the nasopharyngeal neoplasm was removed with the help of semiconductor laser with the wave length of 970 nm in a single block (Fig. 3). There was neither bleeding, nor intra- or postoperative complications.

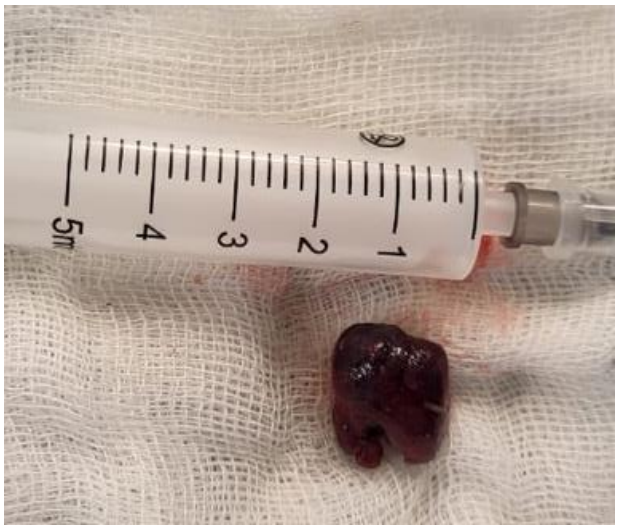

Figure 3. Macroscopic image of the removed nasopharyngeal neoplasm.

The next day after surgery, the child's mother noted a significant improvement in the child's nasal breathing, cessation of dyspnea, and normalization of feeding process. The early postoperative period passed smoothly and the child was discharged to the ENT doctor for outpatient treatment 7 days after surgery.

Histological material obtained for examination demonstrated fragments partially covered with multi-layered squamous epithelium with focal keratinization. A focal growth of granulation high-vascularized tissue of various degrees of maturity with diffuse focal lymphocytic infiltration with a small admixture of neutrophils was found in the superficial parts. The deeper formation has a lobular appearance and biphasic histologic structure: mesenchymal component in the form of intertwining bundles of spindle-shaped cells with round-oval and elongated light nuclei without signs of atypia and mitotic activity, and epithelial component in the form of glandular, trabecular and solid structures consisting of rounded and polygonal cells of glandular type with phenomena of flat cellular metaplasia without atypia, in the lumen of glandular structures positive mucin-exposed secret of various optical density. There were extensive areas of fresh hemorrhages in the stroma with exudative reaction, fibrin deposits, focal petrification, focal fibrosis and mucoidization.

Conclusion: congenital tumor (hamartoma) of the salivary gland rudiment with dyscirculatory changes (fig. 4).

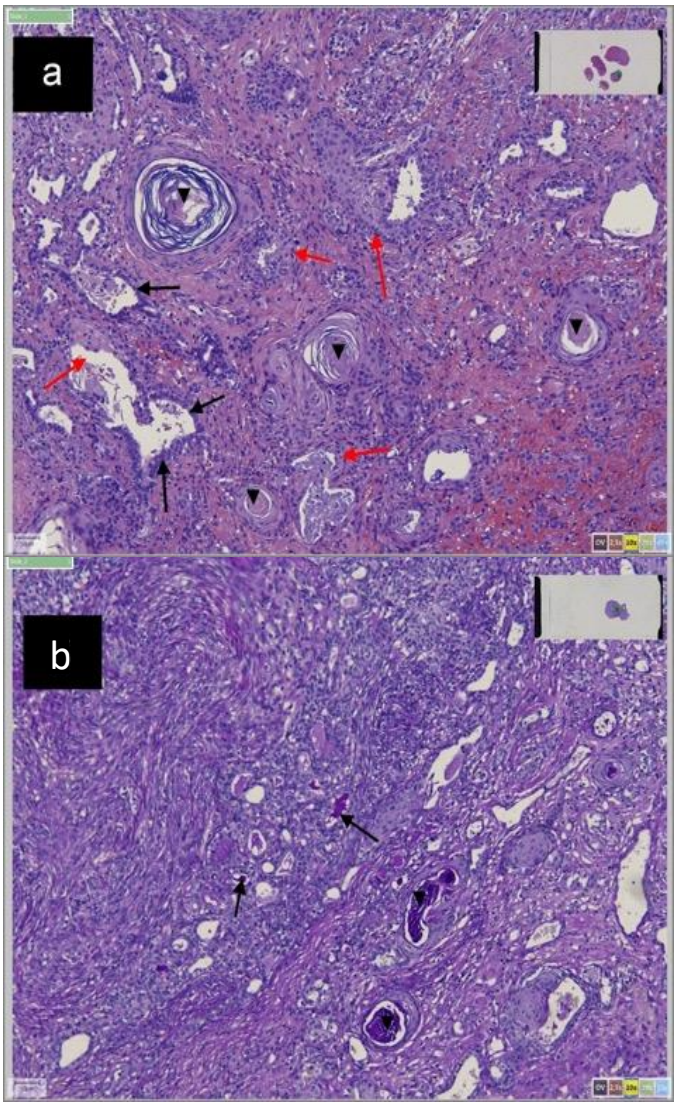

Figure 4. a) Microphotograph shows multiple duct-like structures lined partly by cubic glandular (black arrows), partly by metaplastic multi-layered squamous epithelium (red arrows) with keratinization to the peripheral zones (black triangles). Hematoxylin-eosin staining, x100; b) SHIK-positive kerati-tyne flakes (black triangles) and mucous masses (black arrows) in the lumen of individual dilated ducts can be detected. SHIK reaction, x100.

Control examination of the child 1 month later didn't show any signs of neoplasm recurrence; according to the mother, there were no complaints, the child was active, eating well, there were noticed no signs of apnea, snoring in sleep and obstructive breathing.

Discussion

Nasal breathing disorders in the first 6 months of life can be potentially dangerous as they interfere with feeding, and require careful examination and verification of the cause of nasal obstruction. Respiratory distress syndrome in infants varies widely depending on the area of upper airway obstruction. Volumetric masses in the nasal cavity and nasopharynx in neonates are rare, but their possible presence should always be considered in respiratory distress syndrome in infants during the first months of life [1]. The most common nasal and nasopharyngeal cavity neoplasms are basal cephalocele, nasal cavity glioma (neuroglial heterotopia), and dacryocystocele [2]. Most of these neoplasms in newborn children are benign [3]. Primary and secondary malignant neoplasms of this area are mainly observed in older children, and very rarely

in newborns. They include neuroblastoma, langerhans-cell histiocytosis, rhabdomyosarcoma and lymphoma [4].

The salivary gland anlage tumor (SGAT) in a child presented in our clinical case was first described by Bailie et al. in 1974. [5]. Later, several reports of this neoplasm in children were described in the 1980s and 1990s [6 – 8]. In our country this neoplasm was last time mentioned in publications in 2021 [9]. To date, 42 cases of SGAT have been described in the available publications, and our case is the forty-third one described in the medical literature. Early studies referred to SGAT as a congenital pleomorphic adenoma, suggesting a tumorigenic process [7]. However, later studies deny the tumor nature and present SGAT solely as a benign neoplasm, which dramatically affects its treatment tactics. The absence of destructive changes, morphological similarity to salivary glands, and the absence of recurrence after its radical removal [10 – 12] can prove the benign nature of the process: in the work of Herrmann et al. the postoperative follow-up of a child after removal of SGAT was 5 years [13].

The data analysis shows predominant occurrence of SGAT in males [14, 15]. In most cases, the diagnosis is made in the first months of life, but sometimes SGAT is detected at a later age [16]. Among the leading complaints of children with this neoplasm, prominent is respiratory distress syndrome, i.e., persistent nasal breathing disorders, difficulties in feeding, and slow weight gain. Nasal bleeding is rare. According to the literature, the size of a removed neoplasm varies from 0.5 to 3.5 cm [17].

The main diagnostic methods for suspected nasopharyngeal masses are CT and MRI of the nasal cavity and nasopharynx. These investigations help to determine the connection of a neoplasm with adjacent anatomical structures, absence of intracranial spread, although one case of intracranial spread of SGAT is presented in the literature [18]. Contrast enhancement enables to evaluate blood flow in a neoplasm that is important for planning surgical intervention. Imaging of upper airways is an integral part of diagnostic search for respiratory distress syndrome symptoms, as many specialists focus directly on the evaluation of bronchopulmonary system. For obvious reasons, conservative treatment is ineffective in the case of SGAT.

Endoscopic examination of the nasal cavity and nasopharynx allows visualization of the relation of the nasopharyngeal neoplasm to the surrounding anatomical structures. Thus, the SGAT is characterized by a pedicle location. Total removal of the mass is suggested by the majority of authors (36/42, 85.7%) [15]. Surgery was performed trans-orally, as in our case, or trans-nasally in case of a large mass. Most of the authors did not encounter intense bleeding during the operation.

Histopathologically, SGAT is represented by a combination of benign epithelial and spindle-shaped cells, which form proliferative nodules in the connective tissue stroma. These structures apparently originate from the surface squamous epithelium and are detected in the inter-nodal and per-nodal zones where they merge with stromal-mesenchymal nodules. Some studies have revealed the ultrastructure and immunophenotype of myoepithelium in the latter [19]. In this case, joint expression of epithelial and myogenic immunohistochemical markers confirmed the myoepithelial phenotype of stromal cells. Electron microscopy and immunohistochemistry revealed only epithelial signs in tubular structures and epithelial bands. In this regard, it was suggested that pleomorphic nasopharyngeal adenoma reproduces a salivary gland that develops in the upper respiratory tract [20]. Therefore, in later works, the term "pleomorphic adenoma" was replaced by a “tumor of the salivary gland”. Topographically, the midline location of SGAT is a characteristic feature in comparison with other anomalies in the head and neck region, such as dermoid cyst, nasal glioma, and lingual duct cyst [21]. The fact that no recurrences of SGAT were reported after its removal confirms the benign nature of the neoplasm and its congenital origin (hamartoma).

Conclusions

The described case of the salivary gland anlage tumor in a child is an extremely rare pathology revealed in the first months of life. Symptoms of SGAT are presented predominantly by respiratory distress syndrome, which is a difficulty both in the diagnosis and further management of the patient. Primarily it is conditioned by the age of the patient and nonspecificity of the symptoms: children in the first months of life with symptoms of respiratory distress syndrome are admitted to the pulmonology or infectious diseases department, which in many cases doesn't allow to make a thorough diagnostics and reveal nasopharyngeal neoplasm. Presented clinical case is a typical manifestation of tumor of salivary gland conception - a rare benign nasopharyngeal neoplasm in a child in the first months of life. Early diagnostics including endoscopic examination of the nasopharynx, CT and MRI examinations allowed us to determine the operative treatment tactics, achieve a good functional result in the form of complete relief of

the child's complaints and prevent development of serious complications related to the upper airway obstruction.

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