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A neonate with dyspnea

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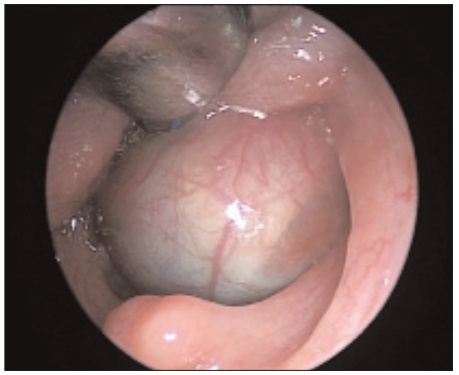
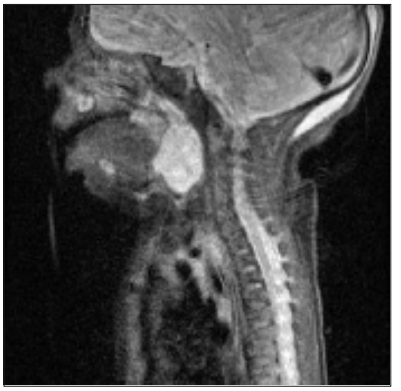
Case report

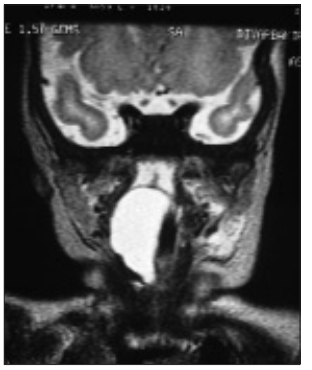
A male baby who was born at term by cesarean section and who had no problem during the prenatal follow-up period was brought to the pediatric emergency department with dyspnea, stridor and gradually increasing cyanosis on the second day after birth. The patient was tried to be stabilized under emergency conditions. Orolaryngeal intubation was performed and the patient was hospitalized in the neonatal intensive care unit. Physical examination including cardiovascular and respiratory systems revealed no pathologic finding. Choanal atresia was absent. Oxygen saturation of patient was 86% in the room air, apical heart rate was 140/min and respiratory rate was 38/min. Blood gases and other hematologic tests were normal. Postero-anterior chest graphy revealed no pathologic finding. Direct laryngoscopy and cervical magnetic resonance imaging (MRI) were performed (Picture 1,2,3).



Picture 1. Direct examination of larynx with laryngoscopy



Picture 2. Cervical MRI



Picture 3. Cervical MRI

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Diagnosis: Mass of the hypopharynx-Hamartoma

Picture 1: Laryngoscopy- a mass occupying the whole hypopharynx.
Picture 2-3: Cervical MRI - a mass occupying the whole hypopharynx

Fiberoptic endoscopic examination revealed a mass occupying the upper respiratory airway to a great extent at the level of hypopharynx. A mass obstructing the airway nearly fully with dimensions of 2x2x2 cm at the level of hypopharynx was observed on cervical MRI (Picture 2 and 3). On direct laryngoscopy, a mass was observed to obstruct the airway fully occupying the lingual root, vallecula and hypopharynx from the level of epiglottis to the level of vocal cords (Picture 1).

When the patient was 12 days old, the mass was extirpated fully using bipolar cautery under direct laryngoscopy. Histopathologic examination confirmed a hamartoma. Our patient is being followed up without any problem and fiberoptic endoscopic examinations revealed no recurrence.

Discussion

Hamartomas are tumor-like formations with a benign characteristic occuring as a result of growing of a form of normal tissue with abnormal arrangement, amount and mixture which may be observed almost in every part of the body (1). It may be defined as an exaggerated growth of normal physiologic tissue which is self-limiting and non-neoplastic and generally occurs in the first two years of life (3). Although it is a benign tissue growth, it may sometimes lead to serious problems locally (3).

In the etiology of hamartomas, causes including congenital malformation, hyperplasia of normal tissue and response to inflammation have been considered (1,3). Hamartomas usually occur as a painless mass and rapid growth may be seen in some cases. Familial characteristics or association with syndromes could not be demonstrated (1,4). Hamartomas can reach a diameter of 15 cm, but they are usually 2.5-5 cm in diameter (3). Differentiating this tumor-like mass from other pathologies including lymphadenomegaly, sarcoma, lipoma, hemangioma, neurofibroma, dermatofibroma or teratoma by detailed investigation is essential in terms of treatment approach (1,3-5).

Hamartomas are usually diagnosed in the neonatal period. They are congenital with a frequency of 15-20% (6). In most cases, the lesion is in the head and neck region. However, it may be found in other body parts and even in regions with no association with skin including the tongue and orbita (7,8). When we screened the literature, we found that no case of hamartoma arising from the hypopharyngeal region has been reported in our country and only a few cases have been reported in the world (5,9). The youngest case in the literature is a 3-year-old patient with a hamartoma arising from vallecula reported by Baarsma (9) in 1979. Our case is the youngest case with a hamartoma arising from the hypopharengeal region who demostrated signs on the second day of life and was operated on the 12th day of life. Spontaneous regression, change or transforming to malignancy have not been reported during the natural course of hamartomas. Lesions which grow rapidly in the beginning start to grow slowly as the age gets older (3). Because of growth in the hypopharynx, hamartoma is one of the rare causes of dyspnea and stridor in newborns among many other causes. In cases who demonstrate no signs, hypopharyngeal hamartoma may be easily overlooked. However, it may cause life-threatening respiratory distress in growing cases. This may be more rapid and severe especially in children. Presence of dyspnea in our case led to early diagnosis.

In these cases, radiologic tests are needed in addition to physical examination. Evaluation with MRI has been reported to be beneficial in the diagnosis of hamartomas (6,10). Histopathologic examination and immunohistochemical studies should be performed to confirm the diagnosis (10). Treatment of this rare pathology is complete excision. However, patency of the respiratory airways should be controlled primarily in the presence of a mass obstructing the upper respiratory airways. Prognosis is rather good. Rate of recurrence is low even after incomplete excision (3).

Consequently, soft-tissue masses in the hypopharyngeal region should be considered among congenital causes in the differential diagnosis when investigating causes leading to dyspnea in the childhood, especially in the neonatal period. It should be kept in mind that congenital hamartomatous lesions may be seen in newborns with dyspnea, though rarely and the cause of dyspnea should be tried to be demonstrated preoperatively by a careful endoscopic examination and radiologic tests if needed (especially MRI). Thus, we can avoid overlooking these cases who show marked improvement with surgical treatment.

Conflict of interest: None declared

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