A Rare Cause of Positional Dyspnea: Hypopharyngeal Hamartoma

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Abstract

Hypopharyngeal hamartomas are quite rare tumors that may cause nutritional or respiratory problems in neonates and infants. They may be confused with other benign laryngeal lesions. In this study a 7-month-old infant with congenital stridor and positional dyspnea caused by a hypopharyngeal hamartoma was reported with review of the literature. Hypopharyngeal hamartomas should be considered in the differential diagnosis of pathologies that cause respiratory or feeding problems in neonates or infants.

Keywords: Hamartoma, dyspnea, hypopharynx, infant

Introduction

Hamartomas are tumor-like masses that contain mature tissue components occuring because of abnormal organization of cells that does not ensure tissue integrity, and grow focally (1). Although rare, hamartomas develop in the head and neck region, sinonasal tract, nasopharynx, oral cavity, oropharynx, larynx, hypopharynx, cervical esophagus, ear, parotid gland, trachea, parathyroid gland, and eye (2). Hypopharyngeal hamartomas are very rare masses that present with asymptomatic or feeding and respiratory problems in neonates or infants. They can be pathologically confused with other laryngeal benign lesions. Hamartomas are usually adequately treated by excision via endolaryngeal microsurgery. However, open surgical interventions are rarely required for large lesions (3). Recurrence is generally observed because of inadequate excision, and even with these inadequate excisions, the rate of recurrence is low. Prognosis is highly good in completely excised lesions (2).

Case Presentation

A 7-month-old infant girl with congenital stridor was admitted to our clinic. Her medical history included respiratory distress that appeared with positional changes. The infant was comfortable in the supine position, but inspiratory stridor occurred when she was in the prone position or left lateral recumbent position. Oropharyngeal examination revealed a smooth-surfaced mass with a vertical movement between the oropharynx and hypopharynx. Flexible fiberoptic laryngeal examination showed a smooth-surfaced polypoid mass that originated from the right posterior pharyngeal wall in the hypopharynx (Figure 1). Magnetic resonance imaging (MRI) revealed a smooth-surfaced lesion that was isointense with muscle tissue and that extended to the epiglottic level at the hypopharynx in the right side (Figure 2). Endoscopic surgery under general anesthesia was planned for the patient. The lesion was completely excised using a diode laser with 30-degree rigid endoscope. The patient did not have any respiratory problems after extubation and was discharged from the hospital after being followed up in the clinic for two days. Histopathological examination revealed that polypoid lesion was covered by mature squamous epithelium and that it had adipose tissue and smooth muscle cells at its core (Figure 3). No mitotic activity was observed in the tissue. The patient was followed up without any problem for one year, and no recurrence was observed in the control examinations performed using fiberoptic...
endoscopy (Figure 4). Written informed consent was obtained from the parents of the patient to use patient’s data and figures in this case report.

**Discussion**

Hamartomas are tumor-like benign formations that can be observed in all body parts and that occur because of the abnormal growth of tissue in an unusual amount and with an unusual mixture of cells (4). While most hamartomas are mesenchymal ones that do not include an epithelial tissue and that develop because of an abnormal growth of the mesenchymal tissue, epithelial-glandular hamartomas, which include a mixture of epithelial and glandular components, are less frequently observed (5). Hamartomas are generally painless and may exhibit a painless
growth pattern. They are congenital, with a rate of 15%-20%, and are usually diagnosed in the neonatal period (6). The association between lesions and familial features or syndromes has not yet been demonstrated (4). As a treatment approach, it is very important to differentiate hamartomas that display a tumor-like growth from structures that form cervical masses such as sarcoma, lipoma, hemangioma, neurofibroma, dermatofibroma, leiomyoma, rhabdomyoma, chondroma, and teratoma (4).

Laryngeal hamartomas may lead to feeding problems, developmental delay, hoarseness, aspiration, and cyanotic spells by causing acute upper respiratory tract obstruction before being diagnosed (7). Similar to laryngeal hamartomas, hypopharyngeal hamartomas are rarely observed. To date, spontaneous regression or malignant transformation has not been encountered in hamartomas. Although the lesion shows a rapid growth pattern at the beginning, the speed of growth decreases with increasing age (8). Compared with other causes of neonatal dyspnea, hypopharyngeal hamartomas are seldom the cause of dyspnea. While hamartomas may be overlooked in some cases owing to the absence of any symptoms, large-sized hamartomas can present with life-threatening respiratory distress. Our case had positional dyspnea that occurred when the hamartoma closed the glottis in the prone and left lateral recumbent positions.

Radiological examination techniques have an important role in the establishment of a diagnosis. In particular, MRI is reportedly useful in the diagnosis of hamartomas (6, 9). Histopathological and immunohistochemical examinations can be used for supporting the diagnosis (9). Total surgical excision is the most important technique for the treatment of these rarely observed pathologies. Hamartomas have a highly good prognosis. The rate of recurrence is very low, even in the case of an inadequate excision (8).

**Conclusion**

Hypopharyngeal hamartoma is a rarely observed soft tissue mass that can cause respiratory distress in infants. Until the appearance of respiratory distress, hamartoma may be overlooked and confused with feeding problems or other pathologies. Endoscopic evaluation and radiological examinations, particularly using MRI, are useful for diagnosis of hamartomas. For the treatment, complete excision of the mass, preferably endoscopically, is adequate.

**Informed Consent:** Written informed consent was obtained from the parents of the patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - İ.K., Y.Ü.; Design - F.Ş., C.O.N.; Supervision - A.V., İ.K.; Resource - C.O.N., F.Ş.; Data Collection and/or Processing - C.O.N., F.Ş.; Analysis and/or Interpretation - İ.K.; Literature Search - C.O.N., F.Ş.; Writing - İ.K., K.K.; Critical Reviews - K.K., İ.K., Y.Ü.

**Conflict of Interest:** No conflict of interest was declared by the authors.

**Financial Disclosure:** The authors declared that this study has received no financial support.

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