Mesenchymal Hamartoma of the Tonsil in a Child: a Case Report

Roberto Campos Meirelles (M.D., Ph.D.)*, Roberto Machado Neves-Pinto (M.D., Ph.D.)** .

* Full Professor of Otorhinolaryngology Department of the State University of Rio de Janeiro - Brazil.
** Lecturer of Otorhinolaryngology Department of the State University of Rio de Janeiro - Brazil.

Addrress to latter: Roberto Campos Meirelles – Rua Siqueira Campos, 43 suite 1125 – Rio de Janeiro – RJ – Brazil – ZC 22031/070 – Phone: 55 21 25485543 – Fax 55 21 25492969 – E-mail: rocame @superig.com.br
Artigo recebido em 20 de março de 2005. Artigo aceito em 18 de abril de 2005.

ABSTRACT

Introduction: The mesenchymal hamartoma of the pharynx is extremely rare and up to now, there are only four cases reported in the current literature.

Objective: To report a case of mesenchymal hamartoma in a child.

Case Report: A 9 year-old child presented with swallowing disorder and eventual suffocation for the last 4 months. A pedunculated tumoral mass was attached to the free surface of her left tonsil and was completely removed under general anesthesia. The tonsil was not removed due to the absence of history of chronic / recurrent infection or suspicion of malignancy.

Conclusions: Mesenchymal hamartomas in children are rare and should be treated by surgical removal.

Key words: hamartoma, oropharynx tumor, tonsillar mesenchymal tumor, mesenchymal tumor, tonsillar tumor, malformation.

INTRODUCTION

Hamartomas are simple and spontaneous growths composed exclusively of components derived from local tissue. The growths produce an excessive number of cells that reach maturity and then cease to reproduce, so that the growths are self-limiting. Hamartomas often present many clinical features of a neoplasm, although they are basically malformations (1).

Hamartomas of the pharynx are rare lesions, mainly in children (Pubmed – National Library of Medicine & LILACS). There are only four cases reported in children (2-4) and six in adults (5-9). In 1979, the first description of this tumor was done, in the oropharynx of a child 3 year-old (2).

Pharyngeal hamartomas can be associated to systemic manifestations or specific syndromes (10-12), among them the Cowden disease (12), a syndrome characterized by macrocephalia, papiledema and hamartomas (11) and the Peutz-Jeghers Syndrome (10).

The purpose of the present paper is the report a case of mesenchymal hamartoma of the tonsil in a 9 year-old child .

CASE REPORT

A female, caucasian, 9 year-old child, came to ORL consultation from pediatrician with the complaints of swallowing disorder with eventual suffocation. No recurrent or chronic tonsillitis. No weight loss. No other symptoms. Duration of the disease: 4 months. Pediatric examination was uneventful. ENT examination revealed a tumoral lesion, red, smooth, pedunculated and attached to the left tonsil, measuring about 2 x 2 x 2cm (Figs 1 & 2). The aspect of the left tonsil was identical to the right one. No infiltration of the pharyngopalatine or glossopharyngeal arches as well as the soft palate. No cervical masses. Then, we chose

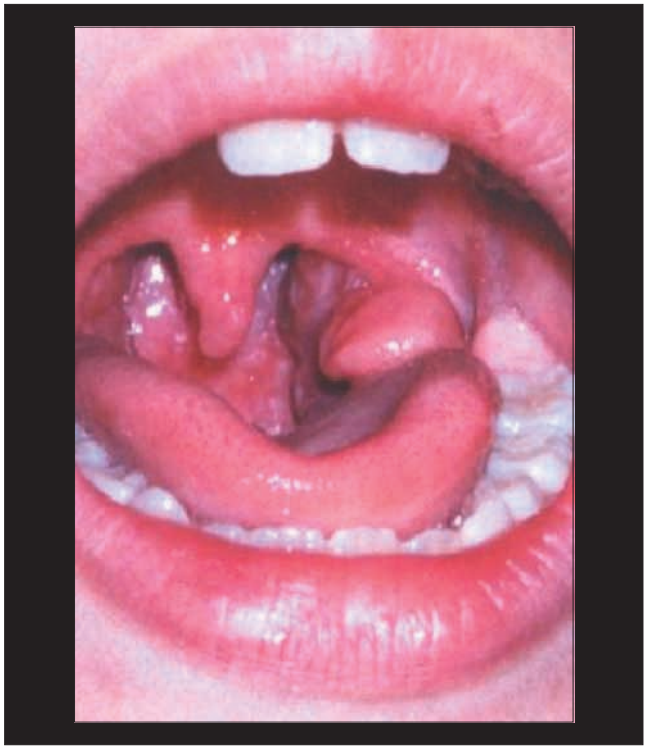

Figure 1. The tumor arising from the left tonsil.

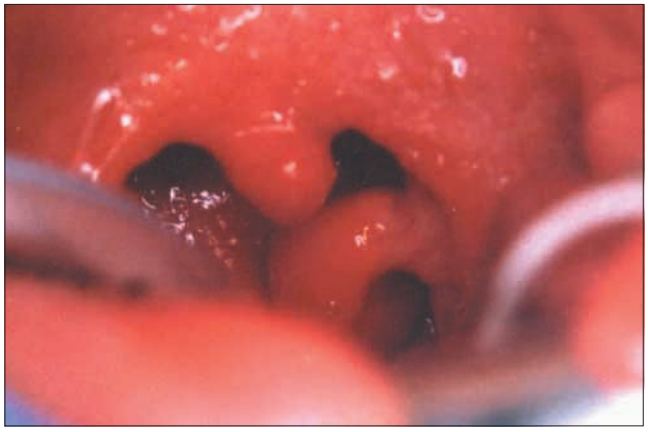

Figure 2. The peduncle of the tumor is clearly seen.

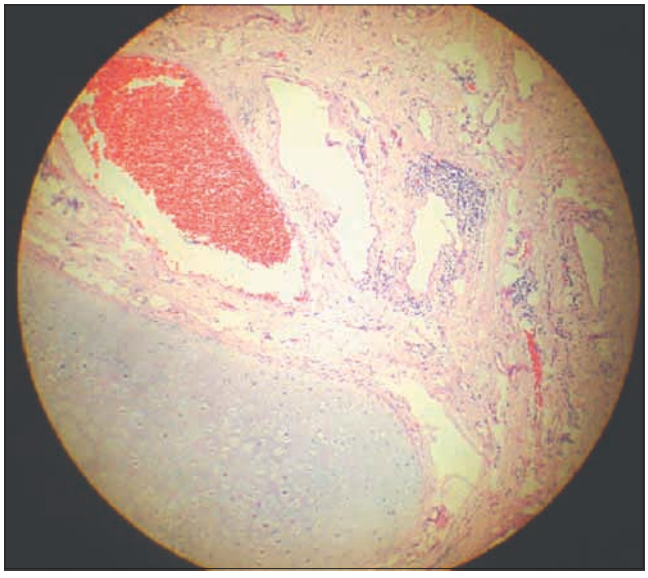

Figure 3. The hysthological picture of the tumor is consistent with the diagnosis of mesenchimal hamartoma.

to perform an excisional biopsy and it was done under general anesthesia, uneventfully. The lesion was completely excised with an additional electrocoagulation of the surgical bed. The tonsil was not removed because it had not a history of chronic or recurrent infection and there wasn´t any suspicion of malignant disease. Surgical pathology diagnosed a tonsillar mesenchymal hamartoma without any suspicion of malignancy (fig 3). The follow up was uneventful.

Discussion

Usually the morphological aspect of the tumor is similar to the one of a polypoid lesion attached to the tonsil, to the wall of the tonsillar bed or to the lateral wall of the hypopharynx, pedunculated, with single or multiple lobulations, smooth surface and a bright red color or pink as the pharyngeal mucosa (9).

Some cases are atypical and can arisen in the pharynx but coming from the anterior or middle cranial fossa with further spreading to the parapharyngeal space. As a rule they present immature brain tissue covered by the meninges (13).

Another possible clinical presentation is the one that comes out in the rhinopharynx ocurring more often in suckling infants, causing feed or respiratory problems. According to the volume of the lesion, asphyxia can occur demanding prompt treatment as an emergency (3,4).

A detailed study of the hamartomas of the pharynx is mandatory in order to differentiate them from tumors like the rhabdomyomas, pleomorphic rhabdomyosarcoma and tumor of the granular cells (7). Some tumors of the tonsils may have two kind of tissue : the lymphoid and the epithelial ones, arising the hypothesis that the polypoids and pedunculated benign tumors of the tonsil could be a mesenchymal hamartoma or branchiogenic remnants only (6). Some of them may contain gastric mucosa (4).

The mesenchymal hamartoma sited in the pharynx is quite rare. In children, diagnosis is usually done before the first clinical manifestation, incidentally, during a routine examination or by occasion of an episode of tonsillitis,

when it can mislead to the suspicion of a malignant disease. Clinical features are dysphagia, snoring and the sensation of foreign body with insidious progression (9). According to the size of the tumor, swallowing disorders, cough, aspiration and eructations can occur (4).

Some of them were found during surgical procedures, when the lesion made intubation difficult. According to its volume the lesion can produce dyspnea. In the case reported by us, the first observation of the lesion was done by the pediatrician who required an ORL examination.

Treatment is through surgical resection. Complete excision of the lesion including its stalk is usually successful. In aldults, surgery can be performed under local anesthesia (6). In the case of voluminous lesions of the pharynx or esophagus and/or in children, general anesthesia is preferred. The tumor is grasped and tractioned to the oral cavity with a forceps for further clamping of its stalk and total excision(8). Even for giant tumors, lateral pharyngotomy was hardly ever necessary (7).

When they are too much vascularized or in contact with cerebral fossae, these tumors can be embolized before surgery through the use of emboli of gelfoam® soaked with an appropriated contrast medium and fluoroscopic control (14).

In the abscence of any extension of the lesion to the bordering structures or communication with the cranial fossa, moreover, being a lesion limited to the tonsillar parenchyma, without invasion of the muscles of the lateral oropharyngeal wall, we chose to perform only the complete excision of the lesion without a tonsillectomy because the tonsil parenchima was looking normal and there was no history of recurrent episodes of tonsillitis. The procedure seemed to be effective as the 4 years follow up showed that the patient remained lesion free and symptomless.

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