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An Abnormal Case of Rhabdomyosarcoma Miming Adenoid Tonsils

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ABSTRACT

Adenoid tonsils are a usual situation in the pediatric population. Their size is the only factor that leads to their surgery. The diagnostic is principally based on clinical symptoms and the endoscopic examination of the cavum. We usually don't need a histopathological evaluation due to the frequency of the situation and also its benignity. The biopsy is needed especially when the evolution is unusual, or abnormal symptoms such as epistaxis or an irregular aspect in the endoscopic evaluation. Our case is about a 3 years old little girl who underwent an adenoidectomy, and 2 months later she developed a malignant tumor in the surgical site.

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Introduction

The hypertrophy of adenoid tonsils is quite a normal situation in pediatric population. We can compare it to serous otitis media. It's an immune reaction secondary to the contact of externa micro-organisms. The problem is once the size of this adenoid tissue is big enough to cause difficulties in respiration and also alimentation. A simple or normal situation can sometimes be the cause of complications such as sleep apnea, facial dysmorphia and others. The surgery is quite easy for the ENT surgeon, but for the anesthesiologist, the upper air ways must be well protected.

It's a benign tumor in most situations, but in rare specific cases we can need a biopsy to make sure if there isn't other tissue. A 3 years old girl who underwent an adenoidectomy, developed an obstruction of the upper air ways in the site of the surgery. The histopathological examination revealed a malignant rhabdomyosarcoma, and that's just 2 months after the surgery.

Case

It's about a 3 years old little girl with a symptomatology of nasal obstruction such as oral respiration, sleep apnea and anterior rhinorrhea. The radiography and the endoscopic examination revealed adenoid vegetation in the cavum.

The patient underwent an adenoidectomy, the adenoid vegetations were removed, confirmed in the histopathological examination. The chest radiography done before the surgery was clean.

The following days the intensity of the symptomatology was reduced, but one week later, it gets worse. One month after the surgery, an adenopathy appeared in the right side of the neck, in the fifth territory related to lymph nodes. The nasal obstruction was complete. The respiration was getting more difficult at the point that we were thinking about making a tracheotomy. In the oral examination, the soft palate was not in its normal position, it as deformed by something pushing it through the oral cavity. (Figure1)

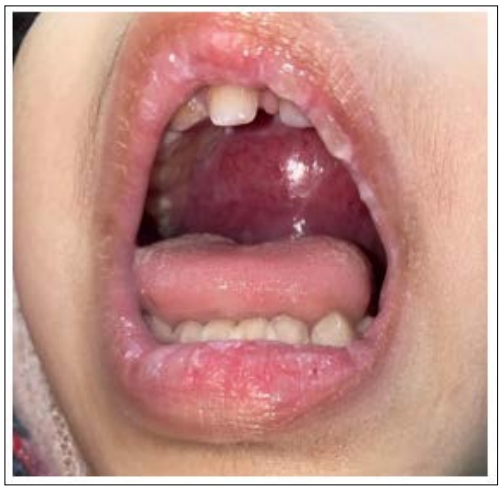

Figure 1: Bulge of the Soft Palate

The CT-Scan showed a mass covering all the space of the cavum, invading a middle skull base, and associated with cervical adenopathy. (Figure 2, 3)

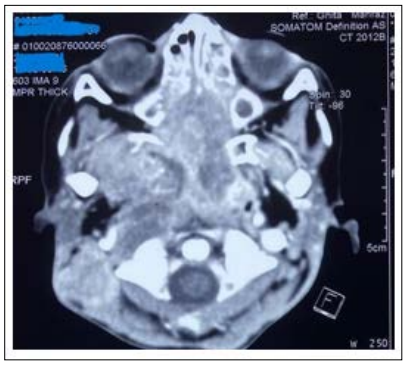

Figure 2: Axial CT Scan Shows the Nasal Cavity and Cavum Totally Filled with a Cervical Adenopathy

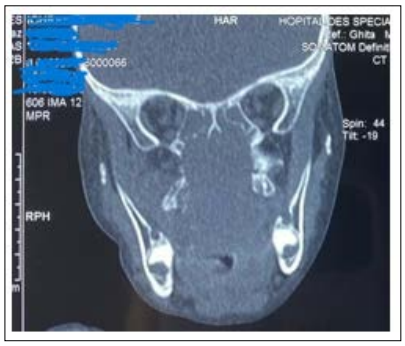

Figure 3: Frontal CT Scan Shows the Lysis of Bony Structures

We have removed the formation that filled the cavum and that took aspect of vegetations especially to free the upper airways.

The histopathological examination of the tissue revealed an embryonal rhabdomyosarcoma. (Figure 4)

Due to the extension of the tumor plus the metastasis, the patient was transferred to oncology department for chemotherapy.

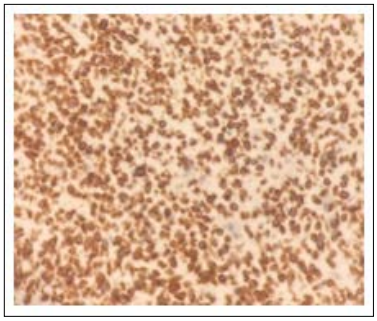

Figure 4: Histopathological Aspect

Discussion

Most of the malignant tumoral processus in the pediatric population are known for their aggressivity unfortunately. Embryonal rhabdomyosarcoma is a malignant tumor derived from primitive mesenchyme [1]. Most of the cases are diagnosed in the pediatric population under the age of 6. The cases are sporadic, and the etiology is still unknown until that day. Some factors are involved, especially the radiation exposure in utero, and also the use of recreational drugs during pregnancy [2]. The problem with this type of tumor is that clinically it can take any form and then could be easily misdiagnosed. Most of the localizations are in the head and neck. From the data of a meta-analysis about 101 patients, the nasopharyngeal localization occupies the fourth position [3]. The nasopharyngeal localization for patients in the age of 6 years or under could be really problematic. At this age, most of the children develop an hypertrophy of adenoid vegetations, and any mass in this situation can be responsible for the same symptomatology as the adenoid vegetations. The nasal endoscopy is much more helpful than a simple radiography to visualize, and at least make a difference between nasal secretions and a mass. However, the embryonal rhabdomyosarcoma can appear in different forms, there is no specific appearance. Our patient had all the clinical characteristics of adenoid vegetations. In fact, the main problems were snoring and oral respiration which initiated the investigations. The endoscopic examination finds out a cavum full of what seems like adenoid vegetations hypertrophy, which is classic in young children. One month right after surgery the symptoms were worse and the respiration were about to be impossible from nasal or either oral airway. The examination finds out the same aspect in the cavum as before the surgery, but this time the size of the mass was important enough to push down the soft palate. We have removed this tissue just to free the upper airways and to avoid a tracheotomy. The tissue was analyzed in the laboratory of anatomopathological where they diagnosed an embryonal Rhabdomyosarcoma. The CT-SCAN was done after the diagnosis, and showed metastasis in lungs and also retropharyngeal and cervical adenopathy. The spread of the tumor was so fast, just one month to develop metastasis. However, the alveolar rhabdomyosarcoma seems to be more aggressive than the embryological rhabdomyosarcoma but the distribution of metastasis does not seem to have any correlation within the first or the second type [4].

Conclusion

The description of that case just shows how a growth of a Rhabdomyosarcoma could be extremely fast and be out of control. All the investigations were done in our case, even the histopathological evaluation of lymphoid tissue, but none could predict the evolution of the situation.

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