



A Rare Case of Laryngeal Angiomyolipoma



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Abstract

Patient's race if known Definition of tuberous sclerosis syndrome (TSP) to differentiate it from AML. A simple drawing of locations would be of value. Is one lesion below the tonsillar area? Are the sizes of the lesion known? Was the surgery done with minimal bleeding? A brief definition of tuberous sclerosis, how it is different from a mesenchymal lesion would enhance this report. A simple diagram of the location of these lesions would add to the value of this presentation.

Introduction

Angiomyolipoma (AML) is a benign mesenchymal neoplasm composed of adipose tissue, blood vessels, and smooth muscle fibres. It is most frequently located in the kidney. The second most common location of the tumor is the liver followed by the abdomen, genital organs, the heart, the mediastinum, lung, skin, head and the neck [1-8]. In the head and neck region, AML has frequently been reported to be in the oral cavity, nasal cavity, and nasopharynx. The median age of incidence is 5th decade. Accompanying tuberous sclerosis syndrome (TSP) is present in 50% of angiomyolipoma patients. AML has been reported in 80% of tuberous sclerosis patients. TSP is defined as, only a few laryngeal AML cases are available in the literature. The lesion is often asymptomatic and usually detected incidentally; AML may present with difficulty in swallowing. Renal AMLs are often invasive, may involve regional nodes and may recur, while on the contrary, AMLs are most often well circumscribed and easily resected. AML seems to follow an entire entirely benign course.

Case Report

We herein report a case of a 57-year old male of Caucasian descent with a left tonsillar region swelling discovered accidentally on routine ENT examination three years ago; The mass itself was pressing on the left lateral pharyngeal space. On video laryngoscopic examination, two separate masses were found- one in the vallecula and a second one in the epiglottic region. Imaging findings supported the clinical suspicion of an AV malformation/haemangioma. Both the masses were totally excised by endolaryngeal microsurgery with minimal bleeding

Location is confusing. First the mass is in the tonsillar region, then they are in the vallecula area. Could we say lower tonsillar region?

Because this is a rare condition, stating the race of the individual would help researchers. If all the cases reported were, for example, if all Asian or other ethnicity, that would be worth knowing.

What are the approximate sizes? Was bleeding minimal?

Grossly, the masses appeared well-circumscribed and globular. On routine haematoxylin and eosin stains, the microscopic examination of the vallecular lesion revealed a well circumscribed lesion formed of thick-walled blood vessels with a few thin-walled capillaries. Some of these vessels are slit-like vascular spaces and some as cavernous spaces. The stroma in between was composed of smooth muscle bundles and lobules of mature adipocytes. Lymphocytic infiltration in the stroma was noted. The epiglottic lesion showed a vascular lesion composed of interconnecting venules and arterioles of varying sizes. Predominant areas of thrombosed and congested blood vessels, haemorrhage and fibrointimal thickening in the venules were noted. The stroma was oedematous. The epiglottic lesion was an A-V malformation. No evidence of dysplasia or malignancy was seen in either of the lesions. Immunohistochemistry done on the vallecular lesion showed positive staining for Smooth muscle actin (SMA) and Desmin by the smooth muscle component of the lesion; whereas CD34 was expressed by the vascular endothelial cells. Immunostaining for melanocytic markers like HMB-45 and Melan-A- which signify a renal angiomyolipoma - was negative. Morphologic and immunohistochemical findings were concordant for the diagnosis of an angiomyolipoma of the vallecula.

Discussion

Angiomyolipoma is a rare benign mesenchymal tumor composed of varying proportions of mature lipid

tissue, smooth muscle fibres, and vessels with thick walls. It is most commonly seen in women. While 50%

of renal angiomyolipoma cases are seen together with tuberous sclerosis syndrome, extra-renal cases are sporadic [9]. There were no additional findings suggesting tuberous sclerosis-

like mental retardation, epilepsy, and cutaneous lesions such as adenoma sebaceum -in our case. Endoscopic preoperative histopathologic diagnosis is difficult due to the sub mucosal development of the tumor. Other tumors of adipose tissue like angioliipoma, liposarcoma and angioleiomyoma should initially be taken into consideration in the microscopic differential diagnosis. Angiolipoma lacks myoid differentiation. Lipoid component does not exist in an angioleiomyoma. In a liposarcoma, lipoid component must be searched carefully for lipoblasts.

Conclusion

The above diagnosis of Angiomyolipoma was made based on both morphology and Immunohistochemistry. The case is being presented for its rarity on presenting within the larynx.

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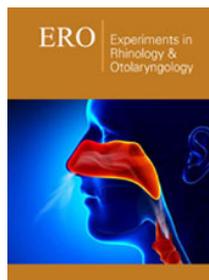
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