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Multicystic Changes of Juvenile Nasopharyngeal Angiofibroma: The First Case Report in the Literature

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***Abstract*— Multicystic changes of juvenile nasopharyngeal angiofibroma: the first case report in the literature. Otolaryngologists, pathologists, and radiologists had better pay attention to this infrequent incidence.**

***Index Terms*- angiofibroma, radiologists, Otolaryngologists.**

I. Case Report

A 13-year-old male patient presented with a 3-month history of right nasal obstruction, epistaxis, mouth breathing at night, and snoring. He was hospitalized in November 2012. Computed tomographic angiography showed that the tumor pedicle originated near the pterygopalatine foramen and extended to the right nasal cavity, nasopharynx, pterygopalatine fossa, and sphenoid sinus (Figure 1, A, B). The tumor was completely removed by surgery. Follow-up more than three years, the patient is in good health.Postoperatively, we found multicystic changes in the peripheral region of the tumor. With the approval of the Institutional Review Board, we herein report this case.

Grossly, the tumor was an irregular, gray-reddish, hollow, triangular mass measuring 6.5 × 4.0 × 3.0 cm. Multiple cystic changes were located in the peripheral region at/or near sphenoid sinus, measuring 0.3 cm to 1.0 cm in diameter. The cross-section revealed that the solid areas of the mass had a gray-white to gray-red and slightly tough appearance, and the cysts had smooth walls (Figure 1 C).

Microscopically, the tumor comprised an intricate mixture of numerous thin-walled blood vessels and fibrous stroma (Figure 2A). The stroma was loose and edematous or myxoid, with spindle or stellate fibroblasts and numerous mast cells and areas of acellular, highly collagenized tissue. The blood vessels, mostly thin-walled venules of variable sizes, had no smooth muscle elastic fibers. Cyst formation was present in some areas. These cysts comprised fibrous walls lined by simple flat cells that partially resembled endothelial cells (Figure 2B, C), which was confirmed by the fact that these lining cells were positive for CD34 (Figure 2D). Immunohistochemically, the stroma cells were positive for androgen receptor and β-catenin.

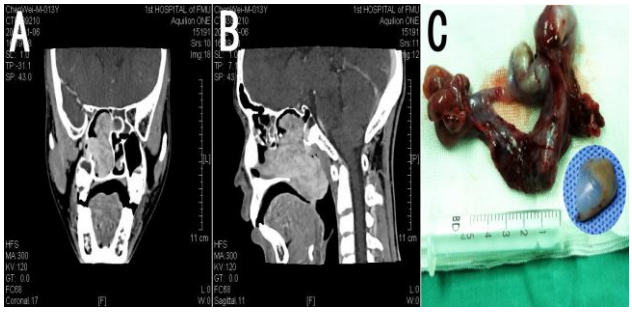

Figure 1: Axial(A) and sagittal (B) computed tomographic angiography demonstrated that the tumor extended into the nasopharynx and sphenoid sinus, but there was a gap between the tumor and the lateral–superior wall of the sphenoid sinus. Resected tumor.(C) with multiple cystic changes in sphenoid sinus region; Inset: A large cyst

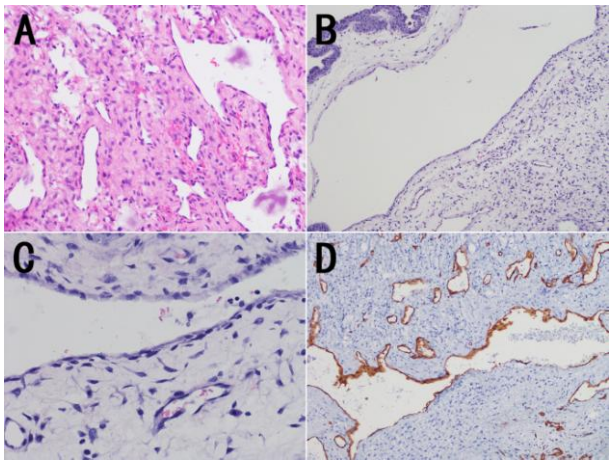

Figure 2: Microscopically, the tumor tissue comprised mainly dense fibrous stroma and numerous thin-walled vessels (A: Hematoxylin and eosin [HE], 200×). The cysts were lined by simple flat cells (B: HE, 100×) that partially resembled endothelial cells under high-power magnification (C: HE, 400×). This was confirmed by the fact that these lining cells were positive for CD34 (D: EnVision, DAB, 200×).

In the present case, cystic changes appeared in the tumor lobule in/or near sphenoid sinus region. A large cyst measuring 1.0 cm contained a small amount of clear liquid.

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Interestingly, the immunostaining findings confirmed that the cells lining the cysts were endothelial cells with a structure similar to that of small, thin-walled vessels.

II. Discussion

To our knowledge, there is no related report in the literature. It is important and interest for clinical and pathological reference. To our knowledge, once a nasopharyngeal angiofibroma has extended to the sphenoid sinus and has become closely adhered to the bony walls of the sphenoid sinus, the cavernous segment of the internal carotid artery often gives off small penetrating branches to this tumor lobule1. In this case, there is a gap between the sphenoid sinus wall and the tumor lobule. Thus, this tumor lobule has no blood supply from small penetrating branches of the internal carotid artery; moreover, it is not located in close proximity to the pedicle region of the tumor. These two conditions are likely the causes of cystic degeneration, and the blood cells are partially or completely destroyed and absorbed; therefore, blood components can not be found in the cysts.

It is well known that cystic changes may occur in kidney and liver2 of human body and in some low-grade brain tumors3. The cystic formation process or mechanism is very complex. This is the first report in which a biopsy-proven juvenile nasopharyngeal angiofibroma with multicystic changes. Otolaryngologists, pathologists, and radiologists had better pay attention to this problem.

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