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CASE PRESENTATION AND LITERATURE REVIEW

Sinonasal smooth muscle cell tumor (Haemangiopericytoma-like tumor)

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ABSTRACT

OBJECTIVE: Haemangiopericytoma-like tumor arising in the sinonasal area is a rare finding in clinical practice. Furthermore, the exact histogenesis of this proliferation is uncertain. Its prognosis is variable, mostly favourable, in the conditions of total surgical removal.

MATERIAL AND METHODS: We present the case of a 64-year-old male with a tumor of the nasal cavity. Routine histological staining and immunohistochemistry were used.

RESULTS: The proliferation was composed of small cells arranged in sheets, and the presence of multiple vascular spaces was obvious. The immunoprofile comprised reactivity for smooth muscle actin and vimentin and, in very rare cells, for CD31 and S100 protein. MIB-1 labeling index was low, about 4%.

CONCLUSIONS: The diagnosis was of haemangiopericytoma-like tumor of the sinonasal area and the patient received no supplementary therapy. Since the tumor is rare, this diagnosis must be acknowledged by surgical pathologists working with otorhynolaringological samples; its histogenesis is not clear and several differential diagnoses are in discussion. Overall prognosis is satisfactory, provided a complete resection is performed.

KEYWORDS: Haemangiopericytoma-like tumor, myoid, glomus tumor, sinonasal, prognosis

INTRODUCTION

Haemangiopericytoma-like intranasal tumor was first described as a particular entity only in 1976, by Compagno and Hyams[1]. Since then, several studies assessed its characteristics, from the clinical and pathological point of view[2,3]. Its nature, although uncertain, seems to be really pericytic according to some authors[4]. However, other authors favor a more close resemblance of this tumor to glomic tumors than to classical haemangiopericytomas[5], from a biological point of view. The tumor occurs in the nasal cavity or sinuses, in middle-aged adults, as a small, polypoid mass[4]. Histologically, the lesion is composed of relatively monomorphic spindled or ovoid cells with eosinophilic cytoplasm and bland nuclei, thus giving the proliferation a rather myoid appearance. The cells are arranged around thin-walled vascular spaces, sometimes taking the staghorn appearance, as in haemangiopericytoma. Mitoses are rare. From the immunohistochemical point of view, most cases exhibit positivity for smooth muscle actin. Vascular markers as CD31, CD34, FVII related antigen are also described in some cases, although with questionable significance and specificity. Its evolution is mostly benign, with only rare examples becoming aggressive (metastatic).

We present a case describing this unusual lesion with emphasis on the pathological and immunohistochemical findings, as well as a brief review of the literature on this topic.

MATERIAL AND METHODS

The patient, a 64-year-old male with a tumor of the nasal cavity, was admitted to an otolaryngology hospital with nasal obstruction. The complete removal of the tumor was performed. The diagnosis was difficult to establish at the local laboratory, and the paraffin block arrived at our Institute for supplementary specifications.

5-μm-thick sections were stained routinely with Haematoxylin-Eosin. Immunohistochemistry was performed using the Envision+ Dual Link System Peroxidase kit (Dako, Carpinteria, CA, USA), ac-

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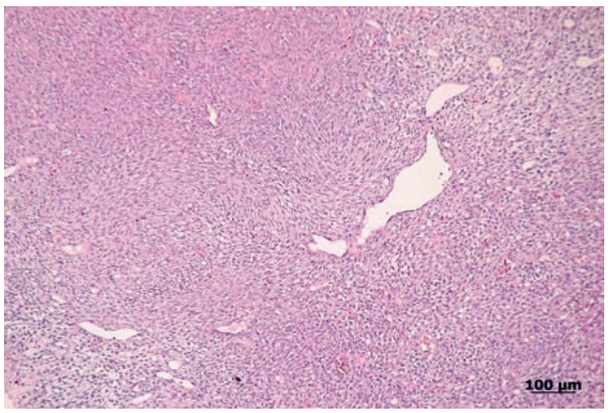

Figure 1 Global aspect of the tumor. The cells are arranged in a patternless manner and vascular spaces are obvious. Haematoxylin-Eosin. Original magnification x100

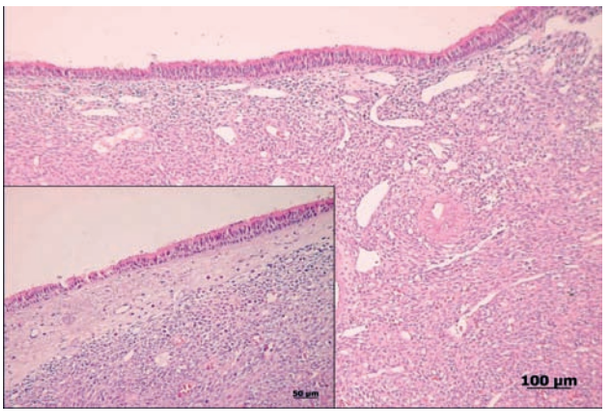

Figure 2 The tumor cells are disposed beneath the epithelium (large image). Insert: a rim of fibrous material separate the epithelium from the main cell mass. Haematoxylin-Eosin. Original magnification x100 (large figure); x200 (inset)

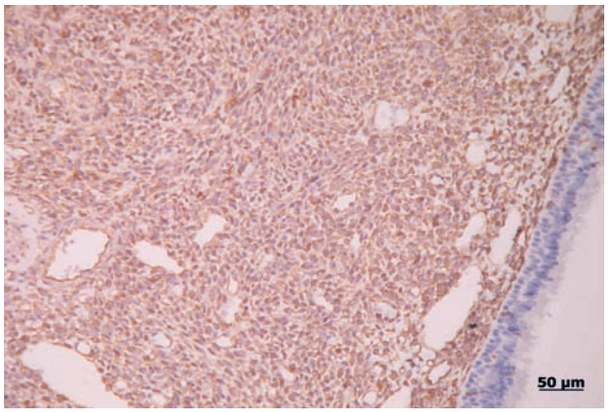

Figure 3 Immunopositivity for vimentin is diffuse in all tumor cells. Original magnification x200

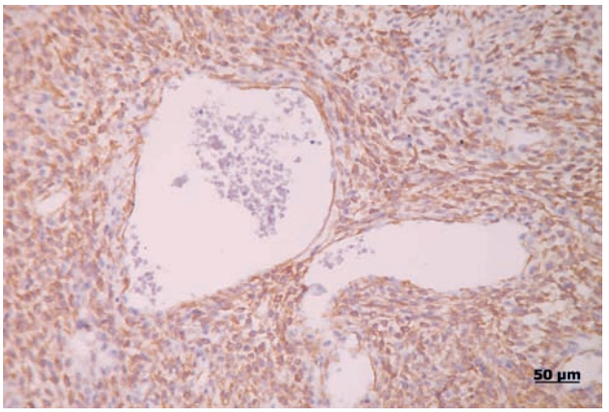

Figure 4 Smooth muscle actin is expressed by all tumor cells. Original magnification x200

cording to manufacturer's instructions. Primary antibodies against the following antigens were used: smooth muscle actin (Dako, Glostrup, Denmark, dilution 1:50), CD31 (Dako, dilution 1:50), CD34 (Dako, dilution 1:50), S100 protein (Dako, dilution 1:50), VIM (Dako, dilution 1:50), CD117 (Dako, 1:250), KL1 cytokeratin (Immuotech, Marseille, France, dilution 1:100), collagen type IV (Biogenex, San Ramon, CA, USA, dilution 1:50), epithelial membrane antigen (Dako, dilution 1:50), MIB-1 (Dako, dilution 1:50).

RESULTS

Routine staining revealed a tumor proliferation composed of oval or spindle cells, with scant cytoplasm, arranged in large sheets in a compact manner, and comprising numerous vascular slits with thin walls (Figure 1). The tumor mass was either immediately close to the epithelium or separated from the latter by a fibrillary area (Figure 2).

Immunohistochemistry revealed a diffuse positivity for vimentin (Figure 3). Smooth muscle actin was strongly expressed by all tumor cells (Figure 4).

CD34 was intense in the vessel walls, but not in the tumor cells (Figure 5). Conversely, CD31 expression was much weaker in some tumor vessels, being slightly positive in rare tumor cells (Figure 6). S100 protein was also found in very rare cells (Figure 7). MIB-1 labeling index was low, not exceeding 4-5% (Figure 8). CD117, KL1 cytokeratin, EMA, and type IV collagen were negative.

DISCUSSIONS

Although sinonasal haemangiopericytoma (SNHP) was thoroughly studied in several papers[2,6-10], its occurrence is still low, at least in otolaryngological practice

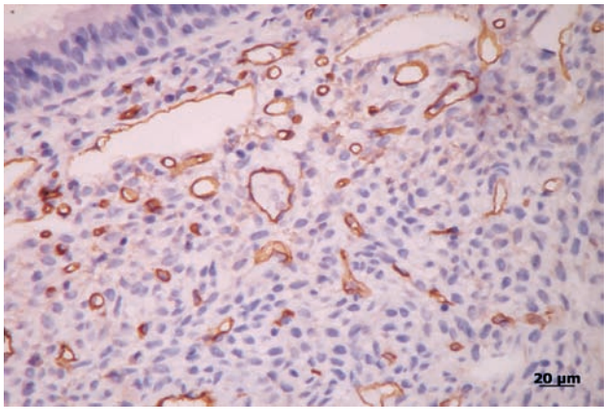

Figure 5 CD34 appears intensely positive in the tumor vessels which were difficult to observe on simple HE staining. The tumor cells are negative. Original magnification x400

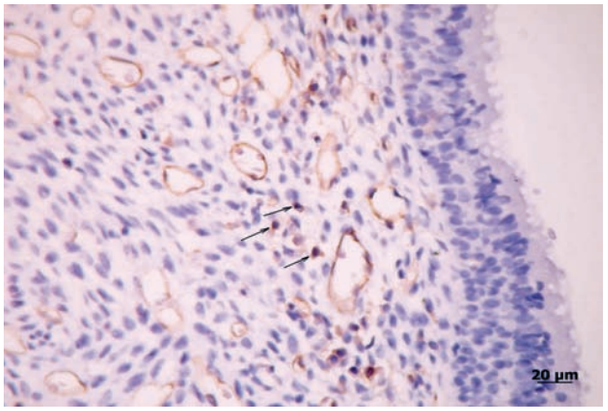

Figure 6 CD34 is positive in vessel walls but also in scattered, rare tumor cells (arrows). Original magnification x400

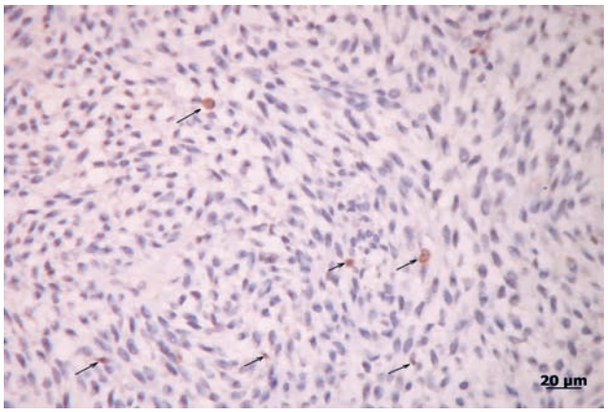

Figure 7 S100 protein expression was restricted to only very rare, scattered cells (arrows). Original magnification x400

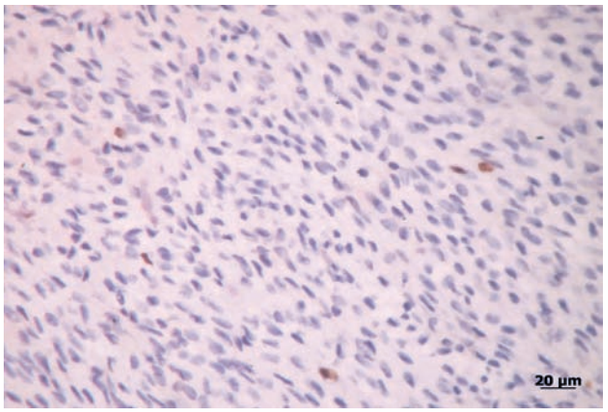

Figure 8 The proliferation index of the tumor is low, below 3%. Ki67 immunohistochemistry; original magnification x400.

and in our experience. Therefore, a characterization of the present case seemed useful mostly for general pathologists not familiar with this particular entity. Its differential also deserves an emphasis in order to avoid misdiagnosis.

The histological examination revealed, in our case, a uniform, diffuse growth pattern. The tumor was located beneath an intact respiratory epithelium. Even though other growth patterns are described (fascicular, storiform, whorled, palisaded, reticular or mixed), they don't seem to influence the overall prognosis of the patient, either regarding the recurrence or the risk of dying of this disease[11]. The same authors describe hyalinized vessels as a quasicharacteristic feature of this tumor type, which was, however, absent in our case. On the other side, the staghorn-shaped vascular channels, specific to soft tissue haemangiopericytoma, were abundant in the tumor stroma. Although some studies emphasized the presence of mast cells in sinonasal haemangiopericytoma tumor, together with eosinophils[11], we did not find their presence, which is in accord with other authors[5]. The mitotic index, constantly found to be low in SNHP[11], was also found at minimal values in our case.

The immunohistochemical reactivity of the tumor was consistent with the diagnosis of SNHP. All tumor cells were reactive to vimentin and smooth muscle actin. This would be in accord with other studies which found a strong positivity for myogenic markers[5]. On the other hand, the presence at least in some cells of endothelial markers as CD31 is not a feature characteristic of glomus tumors, therefore excluding the present case from that possible category of differentials. The lack of tumor cells for CD34 or CD117 also ruled out a potential extradigestive gastrointestinal stromal tumor (GIST). Myogenic differentiation, however, is a characteristic of many smooth muscle tumors arising in the sinonasal region, such as low-grade leiomyosarcoma, smooth muscle tumor of uncertain malignant potential (SMTUMP), and cel-

lular leiomyoma[12]. Anyway, all these entities carry a good prognosis in case of a complete resection. Another proposition for including the SNHP is in a group comprising myofibromatosis, glomangiopericytoma and myopericytoma[13], and defined as "perivascular myomas". Altogether, these tumors have a different histological appearance, with sometimes lower cellularity and no CD31 expression in tumor cells[11], as well as the presence of staghorn vascular slits, very conspicuous in our patient. On the other hand, the meningeal haemangiopericytoma does not express myogenic markers and only inconstantly express CD34. Since our case expressed smooth muscle actin and no CD34 in the tumor cells, it is clear that, despite a histological resemblance between SNHP and soft tissue haemangiopericytomas, it represents a particular entity. The absence of CD34 also excludes a fibrous solitary tumor, characterized by a strong reaction with this antibody, even though CD31 and smooth muscle actin are sometimes reported as positive in this tumor type[14,15]. The lack of epithelial membrane antigen and cytokeratin also excluded a particular form of undifferentiated tumor with epithelial origin. The low proliferation index is in accord with other studies, even though rare cases with aggressive behavior are described, provided a MIB-1 labeling index of >10% is disclosed[11], which was not present in our case.

CONCLUSIONS

Sinonasal haemangiopericytoma is a rare mesenchymal tumor of the nasal region. Its histogenesis remains precisely undetermined, even though a different immunoprofile from its soft tissue counterparts remains to be noted. Its complete removal ensures a good prognosis.

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