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Case Report

Aneurysmal Fibrous Histiocytoma in Nasal Vestibule Mucosa

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Abstract

Aneurysmal fibrous histiocytoma is a different variant of dermatofibroma, characterized by blood-filled spaces within the fibroblastic tumor. In addition to the typical features of a dermatofibroma, it contains large cleft-like or cavernous blood-filled spaces with numerous hemosiderin pigments. This tumor accounts for less than 2% of all fibrous histiocytomas, and it is typically known to occur in the extremities of middle-aged patients. Clinically, they are larger than cutaneous fibrous histiocytomas in size, may be blue, black, or dark red in color, and may have a cystic consistency. They show symptoms of pain and rapid growth. Knowing the characteristics of this rare lesion is important in terms of the differential diagnosis of other malignant mesenchymal neoplasms. In this article, a case of nasal vestibule skin-induced aneurysmal fibrous histiocytoma has been presented in the literature.

Keywords: Vestibule, histiocytoma, fibrous

Introduction

Aneurysmal fibrous histiocytoma is a different variant of dermatofibroma, characterized by blood-filled spaces within the fibroblastic tumor. These tumors constitute less than 2% of all fibrous histiocytomas and are mostly seen in the extremities of middle-aged persons (1, 2). Clinically, they are bigger than cutaneous fibrous histiocytomas, are blue, black, or dark red in color, and may have a cystic consistency. They show symptoms of pain and rapid growth. In 1981, the first case series are reported from Santa-Cruz et al. (3) Knowing the rare features of lesions is important for the differential diagnosis of other malignant mesenchymal neoplasms. Aneurysmal fibrous histiocytomas have the potential to get the wrong diagnosis and differential diagnosis; therefore, malignant melanoma, Kaposi's sarcoma, spindle cell hemangioma, angiosarcoma, and angiomatoid fibrous histiocytoma should be considered (4). Clinically, the aneurysmal fibrous histiocytoma has a tendency to local recurrence more than just an ordinary fibrous histiocytoma; specifically, those histiocytomas that have not been completely excised may have the ability to metastasize, as it is reported (5). This article presents a case of aneurysmal fibrous histiocytoma induced by nasal vestibular skin together with literature.

Case Report

A 67-year-old male patient was admitted to our clinic almost 1 year ago because of the complaints of swelling on the right nasal vestibule. During the anterior rhinoscopy examination of the patient, it was observed that there were approximately 1x1 cm continuous sloping, purple lesions in the right nasal cavity localized to the vestibule (Figures 1a-c). In the patient under general anesthesia, the lesion was excised around a mass of approximately 0.8 mm, and healthy mucosa was left. As a result of the patient's pathology, aneurysmal fibrous histiocytoma was reported. In the microscopic examination of the lesion that was stained with hematoxylin and eosin stain, hemorrhagic, dilated, cavernous vascular channels were observed in the form of slit-like spaces (Figures 2, 3). The patient had no features in postoperative inspections, and we have continued to follow him up. Written informed consent was obtained from the patient who participated in this study.

Discussion

Dermatofibroma (fibrous histiocytoma) is one of the most common types of cutaneous soft tissue lesions (6). It is more frequent in middle-aged adults and has a slight female predominance. Majority of lesions are present as small, raised, hyperkeratotic, cutaneous nodules with a reddish-brown surface (6, 7). A significant part of dermatofibroma is associated with previous minor local

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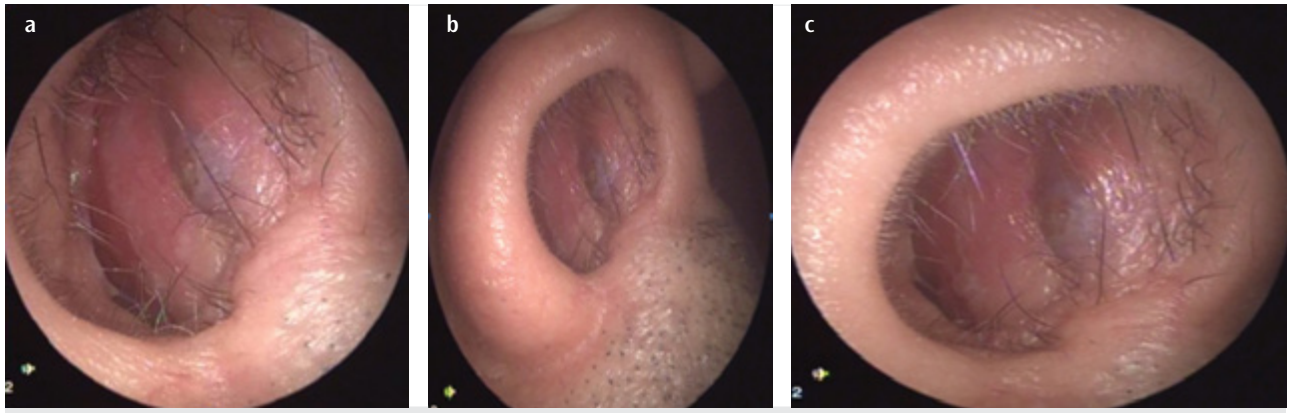

Figure 1. a-c. During the anterior rhinoscopy examination of the patient, there were approximately 1x1 cm continuous sloping, purple lesions in the right nasal cavity localized to the vestibule

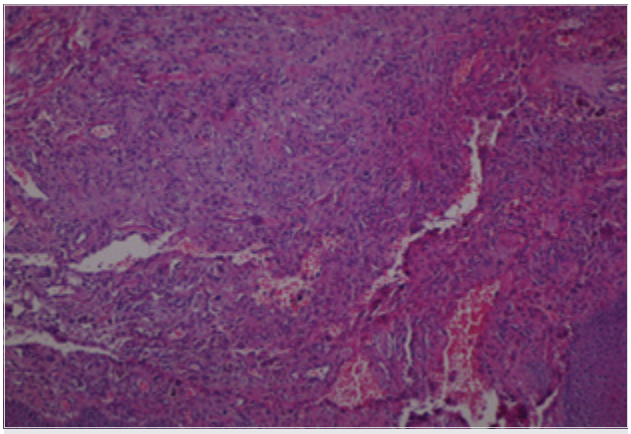

Figure 2. Histopathologic examination under H&E stain revealed hemorrhagic, dilated vascular spaces (H&E stain; original magnification x40)

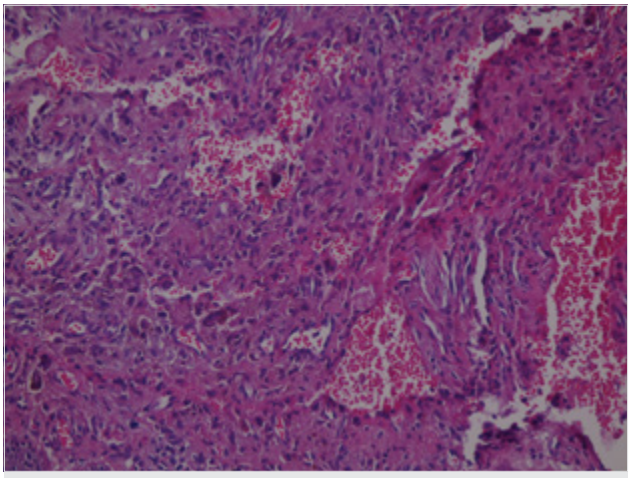

Figure 3. Histopathologic examination under H&E stain revealed hemorrhagic, dilated, cavernous vascular slit-like spaces (H&E stain; original magnification x100)

trauma, particularly insect bites. Eruptive lesions were observed in the context of immunosuppression, HIV infection, highly active antiretroviral therapy (HAART), and pregnancy (8).

Several types of dermatofibroma have been reported in the literature (6, 9). They are essentially distinguished by their histopathological characteristics. However, it is important to note that histological features of several variants can coexist in the same lesion (10). It is important to establish correct diagnosis because some variants also have distinct clinical presentations and different rates of local recurrence. Rare cases of metastasis have also been described (7, 11).

Santa-Cruz et al. (3), were the first who defined "aneurysmal fibrous histiocytoma" as a special variant of cutaneous fibrous histiocytoma. This variant usually represents less than 2% of dermatofibromas and is typically known to occur in the limbs of middle-aged persons (7). In our case, the tumor was localized in the nasal vestibule, as distinct from the cases mentioned in the literature. It has slightly higher recurrence rates (20%) compared with those of other fibrous histiocytomas (1, 2). In our case, recurrences were unidentified during inspections made within the postoperative 6 months, and we have continued to follow him up.

Another tumor, which is similarly called, is "angiomatoid" fibrous histiocytoma, and it is usually located in the subcutaneous tissue. These lesions are described to be surrounded by the lymphoid tissue as a characteristic appearance. These tumors are distinguished from the aneurysmal fibrous histiocytoma with those characteristics (12).

Among the differential diagnosis of aneurysmal fibrosis histiocytoma, malignant melanoma, Kaposi's sarcoma, spindle cell hemangioma, angiosarcoma, and angiomatoid fibrous histiocytoma should be considered (4). The small number of tumor cells in blood-filled cavities formed by the extravasation of red cells may result in the misleading diagnosis of aneurysmal fibrous histiocytoma (1). Previous publications have revealed that the loss of expressions of factor XIII in stromal cells, which surround blood-filled spaces, causes the formation of cavities by disrupting the stromal instability (13). The enlarged spaces become filled with blood, and with the outgoing pressure, they typically consist of cavernous or angiomatoid areas (14). In the microscopic examination of staining with hematoxylin and eosin in our case, hemorrhagic, dilated, cavernous vascular channels resembling slit-like spaces were identified.

As immunohistochemistry, the aneurysmal fibrous histiocytomas are stained negative with the factor XIIIa, CD34, CD31, CD68, and desmine but strongly positive with vimentin (1).

The aneurysmal fibrous histiocytoma clinically tends to local recurrence and has been reported to be even capable of metastatic spreading. Guillou et al. (15), have reported that the greatest risk factors for metastasis are tumor size, necrosis, repeated local recurrence, high cellularity, aneurysmal changes, and a high mitotic activity. During the past years, a few examples of cellular, aneurysmal, atypical dermatofibromas as well as those occurring on the face and in deep soft tissues which metastasized to lymph nodes and the lungs and even caused death of patients in some instances have been reported (16, 17). Dermatofibroma usually show no chromosomal aberrations, but the vast majority of sarcomas typically display a high level of genomic instability manifested by numerous chromosomal aberrations. It has been shown that cases of so-called malignant fibrous histiocytoma (pleomorphic sarcoma not otherwise specified) show multiple genetic aberrations in most of the cases analyzed (18-20), whereas karyotypic alterations in deep fibrous histiocytoma are sporadic and rare (21).

In aneurysmal cutaneous fibrous histiocytomas, knowing characteristics, such as the presence of hemorrhagic pseudocysts, extravasation of red blood cells, and high vascularity, is important to distinguish these tumors between angiomatoid fibrous histiocytomas and cutaneous malignancies of mesenchymal origin (22).

Conclusion

As a rare case, aneurysmal fibrous histiocytomas have the potential to interfere with other malignant mesenchymal neoplasms in the stage of diagnosis and are skin lesions with high recurrence rates, which should be kept in mind.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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