

Case Report

Subconjunctival orbital fat herniation mimicking lipomatous tumors

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Subconjunctival fat herniation is rarely described in the literature, but it is common in clinical practice. It represents prolapse of the intraconal fat to the subconjunctiva, mainly from aging. It usually occurs in the superotemporal quadrant of the globe, and rarely causes symptoms. Microscopically, multinucleated giant cells that frequently have a floret-like appearance (floret-like cells), and also bizarre adipocytes with nuclear vacuoles (Lochkern cells) are seen. We present a case of a 68-year-old Japanese man with subconjunctival fat herniation that showed floret-like cells and Lochkern cells. The differential diagnosis included lipomatous tumors of the orbit such as pleomorphic lipoma, liposarcoma, as these resemble subconjunctival fat herniation macroscopically. It is important to make a histopathological diagnosis for fat herniation to avoid overlooking malignant tumors and over-treating benign lesions.

ACTA MEDICA NAGASAKIENSIA 56: 19 - 22, 2011

Keywords: Orbit, fat herniation, floret-like cell, Lochkern cell, subconjunctiva

Introduction

Orbital fat herniation (orbital fat prolapse) occurs in the subcutaneous region of the eyelid and bulbar subconjunctiva¹. The former situation is common in the elderly, because aging causes extraconal fat to prolapse through the orbital septum. This frequently takes place at the lower eyelid, resulting in so-called "baggy eyelid"¹⁻³. The latter situation is subconjunctival fat herniation (SFH). It usually occurs in the superotemporal quadrant of the globe (bilaterally or unilaterally) by the prolapse of intraconal fat from the weakest point of Tenon's capsule, and is rarely reported in the literature¹. This is also caused by aging or sometimes by trauma^{1,4}. Typically it appears as a yellowish soft mass with superficial vessels¹. On the other hand, since lipomatous tumors resemble SFH in clinical findings and microscopic

features, it is sometimes difficult to distinguish between these two conditions⁵. We report a case of SFH which required pathological differentiation from lipomatous tumors, and review the literature.

Case report

A 68-year-old Japanese man with no ophthalmologic history noticed a small lesion in the left superotemporal conjunctival fornix. It was clinically diagnosed as lipoma, and was followed-up for 3 years. Since the mass had gradually enlarged, the patient decided to have it resected for cosmetic reasons. He was overweight, with a height of 167.5 cm, weight of 79.2 kg, and body mass index (BMI) of 28. He had been treated for 7 years for diabetes mellitus,

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Received December 12, 2010; Accepted February 17, 2011

hypertension, and hyperlipidemia.

Ophthalmologic examination revealed orthophoria, and visual acuity was 20/20 in both eyes. His eye movements were normal and he had no pain or diplopia. Slit lamp biomicroscopy showed a soft yellowish mass located in the superotemporal fornix of the left eye; it measured 15 mm and had a smooth surface and superficial blood vessels (Figure 1). He also had a small white protrusion in his right superotemporal fornix.

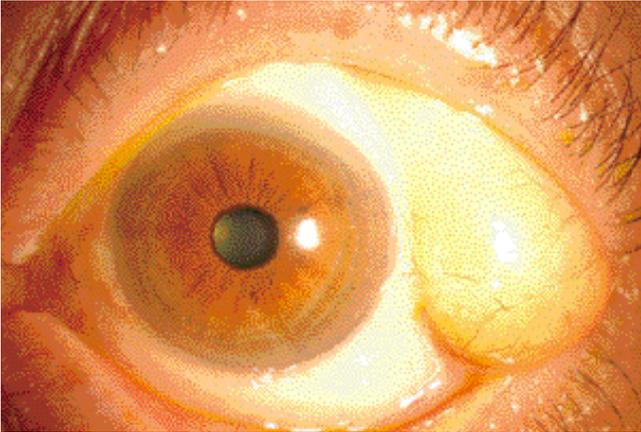


Figure 1. Slit lamp biomicroscopy of the left eye. A soft, well-defined yellow mass is visible in the superotemporal quadrant of the globe. Its surface is smooth and has superficial vessels.

Axial T1-weighted magnetic resonance imaging (MRI) revealed that the mass in the left eye had signal intensity as high as intraconal fat in the lateral portion of the left globe (Figure 2). There was no obvious septum between the lesion and the intraconal fat. These characteristics led to the clinical diagnosis of SFH. MRI revealed no remarkable findings for the right lesion.

Surgical excision of the left mass lesion was decided. Surgical findings showed that the mass was continuous

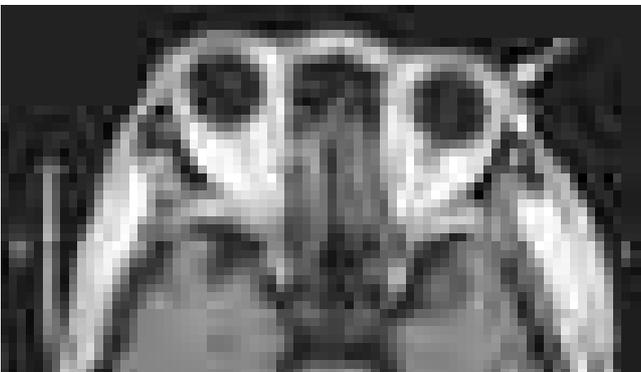


Figure 2. Axial T1-weighted MRI showing a unilateral, high-density mass in the lateral portion of the left globe (arrow). There are no septa between the mass and the intraconal fat.

with the orbital fat, with no evident capsule. We decided on expectant management for the right eye lesion.

One year after the operation, the patient had orthophoria and eye movement was normal as before. There has been no recurrence of herniation in the left eye, and the right eye lesion has not increased in size.

Pathological findings

The specimen was lobulated with fibrous connective tissue. The resected material consisted of mature adipocytes (Figure 3).

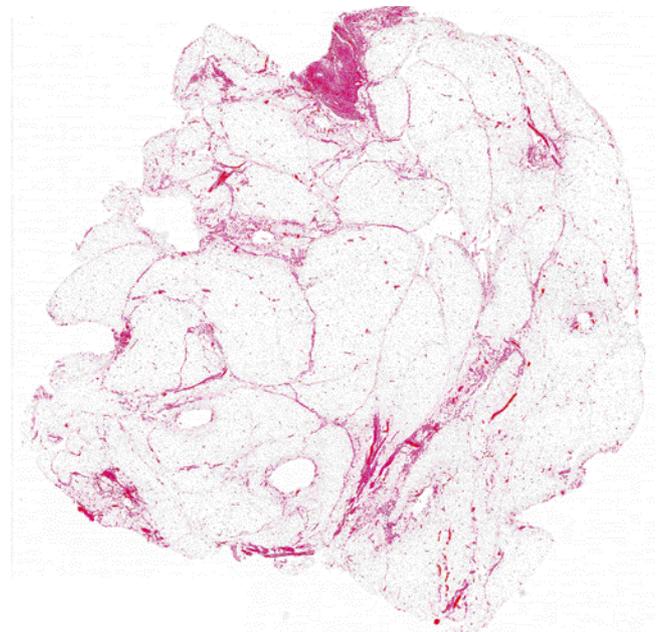


Figure 3. Scanning view of the surgical specimen. The lesion is composed of adipose tissue, lobulated with fibrous connective tissue (hematoxylin-eosin).

Small amount of myxomatous stromal change were located near the fibrous septa. Multinucleated giant cells with the nuclei in a semicircle (floret-like) arrangement and eosinophilic cytoplasm, so called 'floret-like cells', were scattered throughout the lesion. The nuclei of these cells were normochromatic to slightly hyperchromatic, and some showed intranuclear vacuoles. Some adipocytes also demonstrated similar intranuclear vacuoles, consistent with so-called Lochkern cells (Figure 4). In the fibrous area, there were a few eosinophilic collagen fibers and a mild inflammatory infiltrate containing lymphocytes and mast cells.

Immunohistochemically, the floret-like cells, Lochkern cells, and fibroblasts were positive for CD34 (Figure 5).

The pathologic findings suggested pleomorphic lipoma.

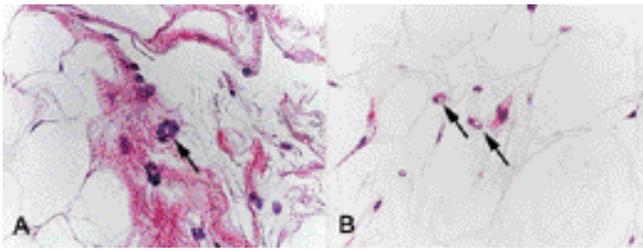


Figure 4. High power view of Floret cells (A) and Lochkern cells (B). The nuclei of these cells are normochromatic or mildly hyperchromatic and demonstrate vacuoles (hematoxylin-eosin x400).

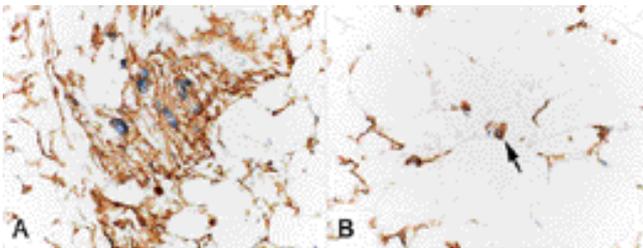


Figure 5. Immunohistochemical findings. Both floret-like cells (A) and Lochkern cells (B, arrow) are positive for CD34 (x400).

However, the findings of mild nuclear hyperchromatism, intranuclear vacuoles of floret-like cells, presence of Lochkern fat cells, and little interstitial myxomatous change, indicated the degenerate nature. In addition, the lesion was not separated from the intraconal adipose tissue by MRI. Therefore, the lesion was diagnosed as SFH, non-neoplastic degenerative lesion.

Discussion

SFH is rarely reported, and its histopathology has been described in only three cases^{1,6,7} in the PubMed literature in the past twenty years (1989 to 2009). It occurs unilaterally or bilaterally, mainly in elderly, obese men. Katayoon et al.⁵ reported 72 cases of SFH, in which 16 (22%) were bilateral, 29 (40%) occurred in men, and 30 (41%) appeared in the superotemporal quadrant of the eye. In bilateral cases, the lesions are generally unequal in size. Therefore minor lesions in the contralateral eye may be sometimes overlooked and the condition diagnosed as unilateral¹. Both SFH and benign lipomatous tumors of the orbit do not usually present with symptoms; however, ptosis or diplopia can occur in rare cases^{1,8,9}. In the present case, the patient was asymptomatic. The mass lesion occurred bilaterally, but the right lesion was very small.

Orbital adipose tissue is normally divided into compart-

ments by fibrous septa¹⁰. Lipoma of the orbit is also composed of lobules of mature adipocytes¹¹, making a definitive diagnosis difficult. Liposarcoma may also be clinically indistinguishable from SFH¹. In addition, the histology of SFH may resemble that of lipomatous tumors, in particular pleomorphic lipoma, myxoid liposarcoma, and well-differentiated liposarcoma. These neoplastic lesions are also reported to occur in the orbital area^{8,9}.

The important differential diagnoses include pleomorphic lipoma and liposarcoma. Floret-like cells are the defining histologic features of pleomorphic lipoma; they are usually located in the adipose tissue and have hyperchromatic nuclei¹. The interstitial myxomatous change is extensive in pleomorphic lipoma¹². Katayoon et al.⁵ reported 72 cases of SFH, in which 31(43%) were found to have floret-like cells. However, Schmack et al.¹ reported that floret-like cells seen in SFH have normochromatic nuclei, frequently present in the fibrous septa, resulting from mechanical irritation by longstanding extraorbital exposure. Moreover, the high power examination of floret-like cells and adipocytes in the SFH frequently demonstrates intranuclear vacuoles. Adipocytes with such vacuoles are reported as Lochkern cells^{1,13}, with *Loch* meaning hole and *Kern* meaning nucleus in German¹³. In the present case, the location of floret-like cells was characteristic of SFH, as were the presence of intranuclear vacuoles and Lochkern cells.

Liposarcoma often presents with proptosis and sometimes diplopia, reduction in visual acuity, and displacement of the globe⁹. Pathologically, necrosis and hemorrhage are often seen in liposarcoma, and there are also multivacuolar neoplastic lipoblasts⁸. In particular, well-differentiated sclerosing liposarcomas can resemble pleomorphic lipoma and SFH, because floret-like cells may occur⁸. So it is difficult to distinguish these entities with precision. Lochkern cells should be distinguished from lipoblasts¹³. Lipoblasts show abnormal-sized and hyperchromatic nuclei with asymmetric shape and mitotic figures. They are also negative for CD34. In contrast, the nuclei of Lochkern cells are normal in size, normochromatic, symmetric in shape, and positive for CD34. Moreover, they do not show mitotic figures.

MRI and surgical findings can also hint at the diagnosis. The mass lesion of SFH is continuous with the intraconal fat, while lipomatous tumors have wispy septations^{1,3}. Lipomatous tumors are visualized as expansive masses with capsules and septa, and there is a recognizable border with the normal surrounding fat.

Because SFH is rarely surgically removed, specimens are rarely examined pathologically. However, microscopy is necessary to perform a differential diagnosis between benign

and malignant lipomatous tumors and SFH. If pathological diagnosis is not performed for this presentation, we may overlook malignant neoplasms that can recur and metastasize to other organs⁹, and we also risk over-treatment of non-neoplastic lesions. Since most of the lipomatous lesions in the orbit are benign, they can be followed up. However, when the lesion is suspected for malignancy, i.e. rapid growth or invasive pattern, the surgical treatment and pathological examination is essential.

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