Intravascular Papillary Endothelial Hyperplasia – A Rare Finding in the Orbital Region

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Key Words

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Abstract

Intravascular papillary endothelial hyperplasia is a benign lesion of vascular origin. It is caused by excessive proliferation of endothelial cells in vascular malformations or normal blood vessels. We report the case of a 58-year-old woman sent to our clinic for surgery of an orbital fat prolapse at her right eye. The clinical examination showed a displaceable swelling with a slightly livid aspect approximately 2 cm in diameter under the upper orbital rim. After MRI, a biopsy was carried out leading to the histological diagnosis of intravascular papillary endothelial hyperplasia. The complete excision of the remaining tumor was performed 8 weeks later. We discuss this clinical entity and the management of such lesions in the orbital region.

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Introduction

Unexplained swellings in the orbital region pose a diagnostic dilemma for clinicians. One of the rarely found entities is intravascular papillary endothelial hyperplasia

(IPEH). IPEH is a benign tumor that mostly occurs as an isolated mass in medium-sized veins without association to systemic diseases. It is considered to be a reactive hyperplasia of endothelial cells developing in response to intravascular thrombosis and thrombus organization [1]. Most of the reported cases in the head and neck region affect the skin and the subcutaneous tissue [2]. The median age of patients with IPEH is reported to be 45 years [3].

The lesions tend to recur if incompletely resected. IPEH mostly occurs in the head, neck and extremities and is rarely found in the orbital region or in the eyelids. We report a case of IPEH in the upper eyelid.

Case Report

A 58-year-old woman was presented for surgical removal of an orbital fat pad. She had been suffering from a slight ptosis and a painless nodular lesion in the right upper eyelid for about 12 months (fig. 1, 2).

Examination of the lid revealed a soft, displaceable, bluish swelling under the orbital rim bulging during Valsalva maneuver. Ocular motility was not affected by the tumor and the patient did not suffer from diplopia at any time. The intraocular clinical findings were unremarkable, the uncorrected visual acuity was 0.63 in both eyes, with +0.5 sph = 0.8.

Magnetic resonance imaging disclosed a 2.5 × 1.5 × 1.5 cm mass in the right medial upper eyelid filling up subcutaneous space completely (fig. 3 and 4). Axial and sagittal T₁- and T₂-

Fig. 1. Nodular lesion in the right upper eyelid bulging during Valsalva maneuver.

Fig. 2. Transpalpebral access showing the vascular tumor.

Fig. 3. Magnetic resonance imaging demonstrating the palpebral nodular tumor.

Fig. 4. MRI showing small coagulations or calcifications within the tumor.

weighted images showed inhomogeneous signals representing small coagulations or calcifications within the tumor. The structure was located above the superior rectus muscle and the bulbous extending approximately up to the bulbous equator without infiltration of the bulbous or orbital structures and without intraconal extension.

The biopsy of this lesion, which was carried out through a transpalpebral access, revealed a dilated vein with a large, partially organized thrombus (fig. 5) surrounded by closely packed myofibroblasts and anastomosing vascular channels harboring plump papillary projections lined by plump endothelial cells. The hyperplastic endothelium showed immunohistochemical positivity for CD31 and displayed no atypia. The diagnosis of an intravascular angiomatosis (IPEH) was made.

Eight weeks later, a complete excision of the IPEH was performed without complications. The patient has been followed-up for 12 weeks without any recurrence.

Discussion

Masson [4] was the first to describe an atypical vascular proliferation, which he named 'vegetant intravascular hemangioendothelioma'. The entity is now called 'intra-

Fig. 5. Microphotograph displaying a dilated vein filled by a thrombus in varying stages of organization being typical for IPEH. Myofibroblasts, vessels with enveloped papillary fronds and hyperplastic endothelium.

vascular papillary endothelial hyperplasia' and has been described in various body sites. The lesion is predominantly situated subcutaneously in the head, neck and extremities, and it is rarely found in the orbital or lid region [5]. Clinically, IPEH appears as a small, firm, superficial mass leaving the overlying skin with a slight red to blue discoloration [6]. IPEH represents approximately 2% of benign and malignant vascular tumors of subcutaneous tissues [7].

IPEH can be found in different vascular proliferations. It predominantly occurs in medium-sized veins but may also arise from pre-existing benign vascular tumors such as pyogenic granuloma, hemangioma and lymphangioma [8].

The diagnosis is based on microscopic examination. The endothelial cell proliferation is histopathologically characterized by confinement to the intravascular space, absence of anaplasia or multiple mitosis, lack of true fibrous papillary fronds and the presence of papillae consisting of fibrohyalinized tissue covered by pleomorphic

endothelium [9]. Since it also shows features like plump endothelial cells, papillary configuration, thrombus formation and irregular blood-filled spaces, it may be histologically mistaken for capillary angiosarcoma.

Masson proposed the lesion to be a primarily endothelial proliferation with secondary thrombus formation [4]. Recent authors consider IPEH to be a reactive hyperplastic process of endothelial cells that develops in response to intravascular thrombosis with subsequent organization of the thrombus [1].

IPEH is known to act aggressively when incompletely resected [1]; still, conservative surgical excision is the treatment of choice. When resected completely, recurrence is extremely rare. In cases where complete surgical removal is impossible, adjuvant radiotherapy or chemotherapy leads to stabilization and short-term regression of the lesion. Gamma Knife radiosurgery seems to be effective in smaller lesions not amenable to complete resection [10].

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