INTRODUCTION Primary histiocytoid carcinoma of the eyelid (PHCE) is a rare tumor, with less than 30 cases described in the English literature (1). PHCE is also named primary signet-ring / histiocytoid carcinoma. It has been also described under different eccrine sweat gland carcinoma names (6) because in the past, PHCE has been claimed to be an eccrine carcinoma. However, some authors argue an apocrine origin and the nature of this lesion is still controversial. It usually affects the eyelid of elderly men and the clinical appearance can simulate inflammatory conditions, which can delay the correct diagnosis. The histopathological exam shows a diffuse infiltrate of small homogeneous cells with histiocitoid and/or signet ring cell appearance. The tumor infiltrates the dermis while the epidermis is usually unremarkable. Immunohistochemical study is helpfull but it is not specific. Therefore, the diagnosis of PHCE is confidently made only after exclusion of a metastasis of carcinoma from other sites. We describe a case of PHCE in a 66 year old man.

CASE REPORT A sixty-six year-old male presented with a history of months progressive painless swelling and ptosis of the left upper eyelid. Patient had no significant past medical or ocular history. At ocular examination, he had visual acuity in both eyes and elevation deficit on the left eye. The palpebral fissure height was mm on the right and mm on the left side. The elevation excursion was mm on the right and mm on the left side. The clinical diagnosis was left blepharoptosis with unexplained poor levator excursion and globe elevation deficit. Acetylcholine antibodies were negative and an orbital magnetic resonance imaging (MRI) was performed. The MRI showed ill-defined soft tissue enhancing lesion on the left upper eyelid. Patient was lost follow-up and six months later he returned with worsening of symptoms. A repeated MRI showed interval increase in lesion size. The lesion has ill-defined borders involving both upper and lower left eyelid. Orbital extension involving intraconal and extraconal spaces with subtotal encasement of the globe was observed. No intracranial involvement or nodal spread was observed. A biopsy of the lesion was performed. Microscopic exam showed tumor composed of monomorphous histiocytoid-type cells with indistinct cell borders arranged between collagen fibers, occasionally forming short files with mild atypia. The tumor diffusely infiltrates the dermis without epidermal involvement. The nuclei were round to oval with finely granular chromatin with occasional nucleoli. Mild to moderate pleomorphism was observed focally. Very rare signet-ring cells were observed. The tumor diffusely infiltrates the dermis without epidermal involvement.
cells and mitoses were observed. Immunohistochemical study showed that the neoplastic cells are positive for CAM\(\beta\), AE1/AE3, CK\(\alpha\), gross cystic disease fluid protein-10, E-cadherin, BER-Ep\(\beta\). The cells are negative for CD\(\alpha\)\(\cdot\)S, \(\alpha\)\(\cdot\)PR, CK\(\alpha\)/\(\cdot\)S and CK\(\alpha\)/\(\cdot\)E. Patient was further investigated for a possible primary tumor or metastasis. No evidence of malignancy was observed elsewhere. The final diagnosis of histiocytoid carcinoma of the eyelid was made. DISCUSSION PHCE is a rare but aggressive tumor. According to the English literature, there are less than 15 cases reported (\(\gamma\)), most of them as single case report. Although this tumor has been described under different names, the debate of an eccrine or apocrine origin is still not concluded and, therefore, we prefer the more morphological descriptive term of PHCE. Characteristically, PHCE affects elderly men with mean age of 57 year-old (ranging from 33 to 78 year-old). The clinical presentation may mimic inflammatory conditions such as chalazion, blepharitis, blepharoconjunctivitis or orbital cellulitis (\(\gamma\)). It can also present as an infiltrative lesion monocle-like appearance (\(\gamma\)). This is important because the biopsy and the diagnosis can be delayed due to this clinical misinterpretation. Histopathologically, it involves the dermis and subcutis of the eyelid without epidermal involvement. The cells can have monomorphic histiocytoid or signet-ring cell appearance or combination of both. Therefore, exclusion of a metastatic carcinoma from breast, gastro-intestinal or other sites is crucial for the diagnosis of PHCE. Although primary breast carcinoma with histiocytoid appearance is typically a variant of lobular carcinoma, lobular carcinoma is almost exclusively a women disease, and it is a well-known tumor that metastasizes to orbit. Therefore, a man with an exclusively eyelid histiocytoid carcinoma, metastatic breast carcinoma may not be considered as a differential diagnosis. However, in a oman, breast should definitely be considered as a possible primary site (\(\gamma\)). At least 0.7% of the cases of PHCE have orbital involvement (\(\gamma\)). There is no immunohistochemical panel exclusively for PHCE. It has been show that PHCE is constantly positive for AE1/AE3, CAM\(\beta\), CK\(\alpha\), gross cystic disease fluid protein-10, BER-Ep\(\beta\) and EMA. It is negative for CK\(\alpha\)/S, CK\(\alpha\)/\(\cdot\)S and S\(\alpha\)/\(\cdot\)PR, Estrogen and Progesterone receptors can be positive (\(\alpha\)), however, these markers are negative in most of the cases. PHCE is known to have a considerable rate of recurrences and metastasis. From 88 cases, 2 patients presented recurrence and 3 patients presented metastasis, 3 of them to the lymph nodes (\(\gamma\)). Metastases to skin, bone marrow, parotid gland and thoracic spine are described, but as single cases. In one case, an aggressive PHCE metastasized to skull, mandible and other bones of the face and vertebrae (\(\gamma\)). Treatment includes wide excision or orbital exenteration with or without radiotherapy. CONCLUSION PHCE is a rare tumor with few cases described in the literature. The differential diagnosis should include metastasis. Immunohistochemistry can be helpful, but a clinical work-up is essential to exclude a primary histiocytoid-like carcinoma from other sites.
این صفحه به معتبر تاییدیه تمامی مقاله در پایگاه استاندارد سیویلیکا می‌باشد. در هر لحظه به منظور تایید اصلاحات این گواهی می‌توانید وضعیت ثبت مقاله را از طریق لینک فوق به صورت آنلاین کنترل نمایید.