Epithelioid hemangioendothelioma of the temporal artery presenting as temporal arteritis: case report and literature review

Dina El Demellawy,1 Ahmed Nasr,2 Salem Alowami3
1University of Northern Ontario School of Medicine, Thunder Bay, William Osler Health Care-Brampton Civic Hospital, Department of Pathology and Laboratory Medicine, Brampton, Ontario, Canada; 2University of Toronto, Department of Surgery, Toronto, Ontario, Canada; 3McMaster University, Department of Pathology and Molecular Medicine, Hamilton, Ontario, Canada

Introduction

In general, vascular tumors encompass a spectrum of tumors with hemangiomas representing the benign group, angiosarcomas that represent the frankly malignant group, and an intermediate group of hemangioendotheliomas. Hemangioendotheliomas are classified as epithelioid hemangioendothelioma (EHE), retiform hemangioendothelioma, composite hemangioendothelioma, Kaposiform hemangioendothelioma (with or without Kasabach-Merritt syndrome), and spindle cell hemangioendothelioma. The latter two types of hemangioendotheliomas usually follow a benign course, in contrast to the other types of hemangioendotheliomas, which are considered as a low-grade malignant sarcoma with unpredictable prognosis. EHEs are rare tumors, mostly described in the lungs and liver. Though endothelial in origin, EHEs reported to originate from arteries are extremely rare. We report a very rare case of EHE arising from the temporal artery showing a peculiar presentation.

Case Report

A 41-year old female presented with painful left temporal artery of one month duration of acute onset and progressive course. The patient’s past medical and family history was insignificant except for bradycardia of unknown etiology and mitral valve prolapse. Physical examination revealed a resting heart rate of 40 beats/min and a thickened palpable left temporal artery. Investigations revealed a sinus rhythm with a rate of 44 beats/min, occasional premature ventricular extrasystole on ECG, mild mitral insufficiency, and mild degree of mitral valve prolapse on echocardiogram. Her cardiac enzymes were negative and chest X-ray was unremarkable. Ultrasound performed on the left temporal artery showed mild aneurysmal dilatation measuring 0.44 cm in maximum diameter. ESR was normal, instead of being elevated as expected in cases of temporal arteritis. Left temporal artery excisional biopsy was performed. Gross examination showed a hemorrhagic vessel measuring 1.2 cm in length and 0.15 cm in diameter that harbored a focal aneurysm measuring 0.5 cm. Microscopic examination showed a solid neoplasm arising from the arterial wall (Figure 1) partially occluding and involving the arterial wall. The tumor was formed of sheets and anastomosing bands of plump epithelioid cells that were embedded in a myxoid stroma (Figure 1). The tumor cells displayed mild pleomorphism, hyperchromatic nuclei, and frequent intracytoplasmic lumina (Figure 2). Mitotic figures accounting for 2/10 HPF were identified, however necrosis was absent. Special stains using Elastic Van-Gieson and Trichrome confirmed that the tumor is originating from the arterial wall (Figures 3, 4 and 5). Immunohistochemistry showed the tumor cells to positively react with CD31 (Figure 6), CD34, Factor VIII and Vimentin and negatively with Cam 5.2, AE1/AE3, EMA, Actin (Figure 7), Desmin,
The case was diagnosed as EHE of the temporal artery. The patient underwent thorough investigation by imaging which documented the absence of any suspicious lesion or residual tumor. The patient was followed up for three years, during which symptoms and signs of recurrence or metastasis were absent.

Discussion

EHE is a distinct entity that was first described by Weiss and Enzinger in 1982, and studied in detail by Ishak et al. in 1984, who described 32 cases. However, a decade and a half later in 1999, Makhlouf et al. reported the largest series of 137 cases. EHE is a rare vascular tumor that is intermediate in morphological features and biological behavior between hemangioma and conventional angiosarcoma. EHE has been reported in the liver, lung, gastrointestinal tract, head and neck, central nervous system, heart, and bone. Tumor demography and presentation is influenced by its location but, in general, these tumors affect middle-aged individuals. Liver and lung lesions are more common in females, whereas tumors of the bone and soft tissues have an equal sex distribution. EHEs present variably with the majority of patients’ symptoms related to the tumor mass effect. Although EHEs originate from endothelial cells, those originating from small sized peripheral arteries are rarely described. To the best of our knowledge, 5 cases of EHEs have been reported (including the current case) (Table 1). However, none originated from the temporal arteries and only a single case showed symptoms mimicking disseminated vasculitis associated with an elevated ESR. In the current case, several differentials were considered including those of endothelial cell origin such as Masson hemangioma (papillary endothelial hyperplasia), epithelioid hemangiomas, and angiosarcoma. The non-endothelial cell origin differentials were metastatic carcinoma, mesenchymal tumors such as intra-vascular fasciitis, myxoid chondrosarcoma and epithelioid sarcoma, metastatic malignant melanoma and pecomas. The presence of the myxoid matrix, cellular atypia, tumor infiltration into the media and adventitia, and the absence of thrombosis excluded hemangioma and papillary endothelial hyperplasia. The absence of profound atypia, brisk mitoses, necrosis and intercommunicating channels excluded the possibility of angiosarcomas and, in addition, the long uncomplicated follow-up of the patient did not suggest angiosarcoma. The immunoprofile of the tumor with diffuse and intense expression of CD31, CD34 and Vimentin, and the absence of Keratin, as well as melanoma, muscle and histiocytic markers is classic for a vascular tumor, excluding tumors of non-endothelial origin. EHE is a well-differentiated endothelial tumor with unpredictable behavior. Unlike angiosarcoma, the histological grading system is not useful for predicting its prognosis. As a result, the treatment options are still controversial. For excised tumors, radiation and interferon therapy may be used in an attempt to restrain growth of incompletely removed tumors. In addition, the former may sclerose the blood vessels. In our case, the patient received no further therapy because thorough imaging showed no evidence of residual disease. In conclusion, we report a remote case of EHE involving the temporal artery and presenting as temporal arteritis. To the best of our knowledge, this is the first case report of EHE in this location and with such presentation.
References


