Atypical fibroxanthoma, rare and often unrecognized cutaneous soft tissue tumor – a case report and review of the literature

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SUMMARY

Atypical fibroxanthoma (AFX) is a rare cutaneous soft tissue tumor typically occurring in the elderly on sun exposed skin. Histologically, it is composed of pleomorphic, atypical cells with multiple mitoses including atypical mitotic figures resembling undifferentiated malignant tumor. AFX is considered to be a benign tumor with almost uniformly excellent prognosis following conservative therapy if strict diagnostic criteria are applied. We present a case report of 68-year-old man with a skin tumor in the temporo-parietal region. Histomorphological and immunohistochemical analysis led us to the diagnosis of atypical fibroxanthoma. We offer a review of terminology and categorization of this tumor and an overview of immunohistochemical markers useful in differential-diagnostic process to rule out other malignant tumors, because AFX is a diagnosis of exclusion. The correct diagnosis prevents unnecessary overtreatment of the patient.

Keywords: Atypical fibroxanthoma – undifferentiated pleomorphic sarcoma – immunohistochemistry.

Atypický fibroxantóm, zriedkavý a často nerozpoznaný kožný mäkko-tkanivový nádor – kazuistika a prehľad literatúry

SÚHRN

Atypický fibroxantóm (AFX) je zriedkavý kožný mäkko-tkanivový nádor typicky sa vyskytujúci u starších ľudí na koži vystavenej slnečnému žiareniu. Histologicky je tvorený pleomorfnými atypickými bunkami s početnými mitózami vrátane atypických mitóz, čím pripomína nediferencovaný malígny nádor. AFX je považovaný za benígny nádor s takmer výlučne excelentnou prognózou po konzervatívnej liečbe, ak sú použité prísne diagnostické kritériá. Prezentujeme prípad 68-ročného muža s nádorom kože v temporo-parietálnej oblasti. Histomorfologické a imunohistochemické nálezy zodpovedali diagnóze atypického fibroxantómu. Ponúkame prehľad terminológie a kategorizácie tohto nádoru a spektrum imunohistochemických markerov nápomocných v diferenciálne-diagnostickom procese na vylúčenie iných malígnych nádorov, pretože AFX je diagnóza *per exclusionem*. Správnou diagnózou sa vyhneme nepotrebnej liečbe pacienta.

Kľúčové slová: Atypický fibroxantóm – nediferencovaný pleomorfný sarkóm – imunohistochémia.

Cesk Patol 2019; 55(3): 182–186

Atypical fibroxanthoma (AFX) is a rare cutaneous soft tissue tumor typically occurring in the elderly on sun exposed skin. It comprises up to 0,2% of all skin tumors (1). AFX is considered to be benign tumor with excellent prognosis after complete surgical excision with free margins. However, the histological structure of AFX remarkably resembles an undifferentiated malignant tumor, that needs to be excluded in differential diagnostic process. Incorrect diagnosis may lead to unnecessary overtreatment of the patient.

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CLINICAL HISTORY

68-year-old man with a skin tumor in the right temporo-parietal region was referred to the plastic surgeon by a dermatologist with the diagnosis of haemangioma with recorded trauma in that region in the past. Plastic surgeon´s clinical diagnosis was suspicious pyogenic granuloma.

MATERIALS AND METHODS

Formalin-fixed, paraffin-embedded tissue blocks were cut into 5-µm sections, stained with hematoxylin-eosin and analyzed immunohistochemically with different primary antibodies: AE1/3 (clone AE1/AE3, diluted 1:300, DAKO, Denmark), vimentin (clone V9, prediluted, DAKO, Denmark), SMA (smooth muscle actin, clone 1A4, diluted 1:200, DAKO, Denmark), desmin (clone D33, diluted 1:100, DAKO, Denmark), h-caldesmon (clone h-CD, diluted 1:100, DAKO, Denmark), beta-catenin (clone β-catenin-1, prediluted, DAKO, Denmark), S100 (clone Anti-S100, diluted 1:1000, DAKO, Denmark), Melan-A (clone A103, diluted 1:200, DAKO, Denmark), HMB45 (clone HMB45,