Epithelioid hemangioendothelioma presenting as fever of unknown origin

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Epithelioid hemangioendothelioma (EHE) was first described by Weiss and Enzinger in 1982 and has been suggested to be a malignant vascular neoplasm with indolent behavior [1]. A 65-year-old female patient presented to our hospital with fever, back pain and pain in her buttocks. On examination, the only pathological sign were cutaneous nodules appearing in her right thigh (Figure 1 A). The histopathology of the nodule was compatible with epithelioid hemangioendothelioma (Figure 1 B). Positron emission tomography/computed tomography (PET/CT) scan revealed the presence of multiple lesions in various muscle groups as well as the invasion of bone marrow. The patient was referred to the oncology department, where she was treated for an angiosarcoma, with the combination of gemcitabine with docetaxel.

Epithelioid hemangioendothelioma is a vascular neoplasm of endothelial origin, which is considered to be intermediate between hemangioma and angiosarcoma [2]. It usually presents as a solitary, slightly painful papule, nodule or non-healing ulcer. Rarely, it may present as multiple lesions [3]. Epithelioid hemangioendothelioma is CD31, ERG, Fli-1 and factor WIII positive [4]. Occasionally, CD30 expression, although mainly associated with lymphoid malignancies and germ cell tumors, has also been reported in EHEs. This is important for the differential diagnosis, but it may also have therapeutic implications as CD30 positive patients could be candidates for targeted therapy based on the new CD30 antibody agents [5]. In most cases, a definite diagnosis of EHE remains challenging. Chromosomal translocation t(1;3)(p36;q25) is characteristic for EHE, and may serve as a diagnostic tool [1].

To our knowledge, this is the first report of EHE which presented as fever of unknown origin, due to its spread in the bone marrow, a fact that became prominent on PET/CT scan. Thus, this case also highlights the usefulness of PET/CT scan in the diagnostic algorithm of fever of unknown origin.

Conflict of interest

The authors declare no conflict of interest.

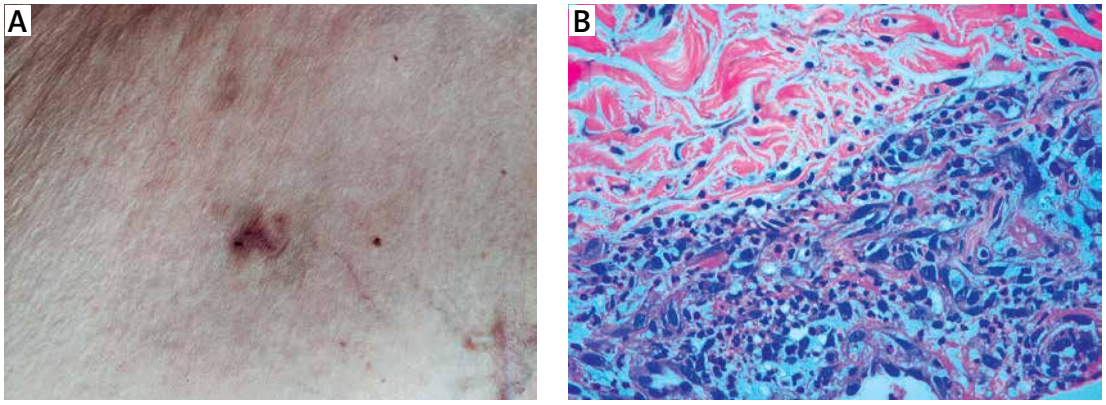

Figure 1. A – One of the cutaneous lesions that became visible and were compatible with epithelioid hemangioendothelioma. **B** – Hematoxylin and eosin 400×: Tumor consisted of a mixture of slightly pleomorphic spindle and epithelioid cells with abundant, sharply defined eosinophilic cytoplasm and vesicular nuclei containing single nucleoli

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