LETTER TO THE EDITOR

A case of inflammatory myofibroblastic tumour of the palm in a young girl

Editor

Inflammatory myofibroblastic tumour (IMT) is a distinct tumour entity of soft tissues first defined in the 1994 World Health Organization classification of tumours of soft tissue and bones. It is described as a lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes and eosinophils.[1] IMT often affects young adults and all body sites can be involved.

Only few cases of IMT have been previously described in the skin.[2] Because of its abundant inflammatory infiltrate IMT has being referred also as inflammatory pseudotumours (IP). IMT is microscopically similar to plasma cell granuloma (PCG) but they are now considered unrelated entities because of a completely different clinical setting.[3]

Herein, we report an additional case of IMT occurring in a 14-year-old caucasian girl who presented to the dermatological clinic because of an asymptomatic subcutaneous, slowly growing, 4-month-old nodular lesion on her right palm. The overlying skin was normal (Fig. 1a). No trauma, surgery or insect bite was reported in that site. Physical examination was otherwise normal, routine serum and urinary examinations were within normal ranges.

The lesion was surgically removed; it was a spherical, well-defined, nodule with a firm consistency and a greyish colour located in the superficial dermis and easily removable (Fig. 1b inset).

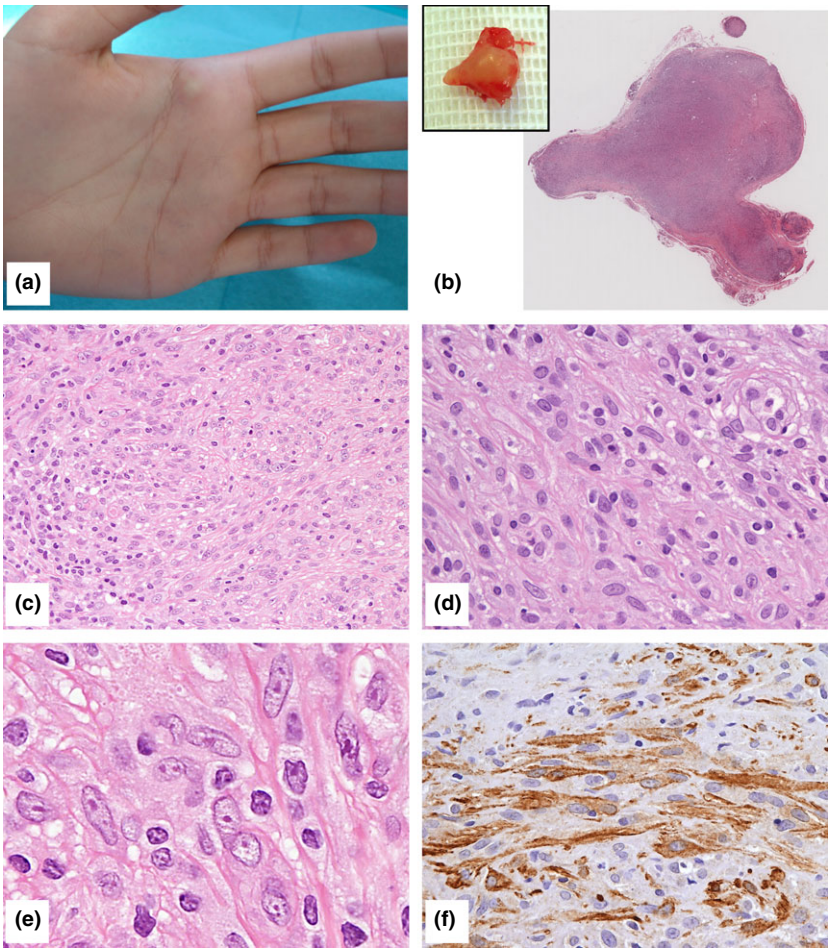

Figure 1 The tumour presented as a well-defined subcutaneous lesion of the left palm (a). After resection a spherical, capsulated nodule was evident with a firm consistency and a greyish colour (b inset). On histology, at low power, (b) the lesion was highly cellulated and delimited by a thin fibrous capsule (Haematoxylin and Eosin, scan image by Aperio Scanner). At higher power the tumour showed proliferation of spindle cells with plump ovoid, nuclei, with prominent single or multiple nucleoli, intermingled with reactive inflammatory infiltrate mainly made of small lymphocytes (c, d and e; Haematoxylin and Eosin 200×, 400×, 500×). By immunohistochemistry spindle cells showed expression of smooth muscle actin (f; 200x).

On histology, a thin fibrotic capsule delimitated a nodular proliferation (Fig. 1b) made by spindle-plump cells with oval or roundish nuclei, single or numerous prominent nucleoli and abundant eosinophilic cytoplasm intermingled with a polymorphic lympho-plasmacytic infiltrate (Fig. 1c–e). Very scant interposed stroma was present. By immunohistochemistry, spindle-plump cells showed partial expression of the smooth muscle actin and were negative for melanocytic and vascular markers (S-100 and CD34 respectively). (Fig. 1f) HHV8 infection was excluded (HHV8/ORF73 antibody). ALK protein (ALK/p80 Ab-1 antibody) was not expressed by tumour cells.

Only 19 cases of cutaneous inflammatory myofibroblastic tumour (cIMT) have been previously described in the English literature,[2,4,5] none on the hand palm. Clinical presentation is reported as rather aspecific, cIMT develop as deep single nodules of hard consistency and occasionally appear as exophytic ulcerated tumours.[4,6] Lesions are always reported as asymptomatic with a benign course.

On histology, IMT should be differentiated from spindle cell neoplasia such as dermatofibrosarcoma protuberans and solitary fibrous tumour; melanocytic proliferations with abundant inflammatory infiltrate should also be excluded.[2]

Inflammatory myofibroblastic tumour is a clonal proliferation of myofibroblastic cells, as demonstrated by cytogenetic studies[7] and translocations involving ALK (anaplastic lymphoma kinase) gene are detectable in about half of the cases, if considering all body sites.[8] ALK gene can translocate with CARS, CLTC, RANBP2, ATIC, SEC31L1, TPM3 and TPM4, resulting in different patterns of ALK protein expression by immunohistochemistry.[9]

In all body sites other than skin, the lack of ALK protein expression is correlated with an adverse prognosis and it generally occurs in older patients.[10] This is not true in cIMT since all reported cases are ALK negative and all showed a benign clinical course.[4]

This could suggests a different, still unknown, genetic abnormality responsible for IMT development in the skin; since HHV8 infection was reported in a single case of cIMT, a clonal proliferation triggered by an infectious event could not be excluded.[6]

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