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CASE REPORT

Case report of a clinically indolent but morphologically high-grade cutaneous mast cell tumor in an adult: Atypical cutaneous mastocytoma or mast cell sarcoma?

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Abstract

We present a case of an adult male with a solitary mast cell tumor of the skin with unusual nuclear pleomorphism and mitotic activity. The tumor was excised, recurred within 2 years, was reexcised after 4 years and did not recur >6 years after diagnosis. The tumor showed progressive cytonuclear atypia and a high mitotic and proliferation rate by Ki67-staining from the onset. No *KIT* mutations were identified in the tumor and bone marrow. Serum tryptase levels and a bone marrow aspirate and trephine biopsy were normal. Although the histomorphology of the skin tumor was consistent with mast cell sarcoma, the clinical behavior without systemic progression argued against this diagnosis. The tumor was finally considered as atypical mastocytoma, borderline to mast cell sarcoma. Currently, the patient is in close follow-up and still in complete remission.

KEYWORDS

adult, case reports, mast-cell sarcoma, mastocytoma, mastocytosis

1 | INTRODUCTION

Mastocytosis is a heterogeneous disease, characterized by expansion and accumulation of clonal mast cells. The disease may be limited to the skin (cutaneous mastocytosis), may be systemic (indolent, smoldering, aggressive, or leukemic) or rarely presents as a malignant solid tumor with destructive growth, known as mast cell sarcoma (MCS).^{1,2}

Localized cutaneous mastocytoma mainly presents in prepubertal children, but is extremely rare in adults.^{1,3} The exact incidence is unknown.³ In the literature, only 14 cases of adult purely cutaneous mastocytomas have been reported.⁴⁻¹⁵

In cutaneous mastocytosis and indolent systemic mastocytosis, mast cells have round or oval nuclei and abundant metachromatic cytoplasm,¹ or are spindle-shaped hypogranulated cells being referred

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as atypical mast cells type I.^{1,16} A third type of cytology consists of mast cells with nuclear pleomorphism, including bilobation and/or multilobation; these cells are known as atypical mast cell type II or promastocytes.¹⁶

In the literature, nine cases of benign mastocytosis or mastocytoma of the skin with type II atypia have been reported,^{7,17-21} including two adult cases.^{7,17} Although histomorphology raised concern about a malignant behavior, none of these patients showed systemic involvement or destructive growth, and after excision no recurrence was observed.⁴⁻¹⁵

Here, we report on an atypical solitary mast cell tumor in a middle-aged patient, which in contrast to the other described cases recurred after excision, showed progressive cytological pleomorphism and an unusually high proliferation rate, but did not show any signs of clinical malignancy after more than 6 years of follow-up.

2 | CASE REPORT AND RESULTS

2.1 | Case report

In January 2014, a 44-year-old male presented with a solitary yellowish plaque on the skin of the right temple (Figure 1A, Table 1). The lesion had been stable for 5 months and was itchy upon touching. His case history reported a thin superficial spreading melanoma on the right upper leg in 2012 (the diagnosis being confirmed upon revision). There was no history of pediatric cutaneous mastocytosis.

Histopathological examination of a punch biopsy showed diffuse sheets of epithelioid cells in the dermis, without involvement of the epidermis or skin adnexa. The cells had abundant, pale eosinophilic, ashy to finely granulated cytoplasm, and contained no melanin pigment. The nuclei were slightly polymorphic, with mostly round and some oval forms. Spindle-shaped cells, and cells with irregularly shaped, bilobed and multilobed nuclei were sparse. There was a low, nonatypical mitotic activity, with up to one mitotic figure/mm². The Giemsa-stain highlighted the metachromatic granules and scant intermixed eosinophils. In this and later biopsies, immunohistochemistry was positive for CD117, tryptase (Figure 2B), and microphthalmia-associated transcription factor.

Other neoplastic conditions were excluded by immunohistochemistry using antibodies against S-100, Melan-A, HMB-45, SOX-10, pan keratin (CKAE1/3), p40, CD1a, and CD34. The CD30 stain showed a heterogeneous pattern with some strongly positive tumor cells. In the CD25-stain, there were a few positive cells. Neoplastic cells stained negative for CD2. Next generation sequencing analysis did not show mutations in *KIT* (including exon 17).

Physical examination disclosed normal results. Laboratory testing showed a normal serum tryptase level (2.9 µg/L, normal value <11.4 µg/L). No further staging procedures were performed.

An excision followed in July 2014. Histopathology revealed a similar picture compared to January 2014, with a slight increase of pleomorphic nuclei and mitotic activity (Figure 2A). In a hot spot, up to nine mitotic figures/mm² were found (Figure 2C). The Ki-67 proliferation index reached up to 29.5%. In addition to the well demarcated tumor bulk, the CD117 and tryptase highlighted perivascular and periadnexal continuation of small clusters of mast cells, including few pleomorphic mast cells, into the resection margins (Figure 2D).

2.2 | Provisional diagnosis

Based on histopathological studies and staging investigations, a provisional diagnosis of a mastocytoma was made, because of the clinical presentation of a solitary nodule without evidence of systemic disease, non-destructive growth and the low proportion of atypia.

2.3 | Clinical course in the follow-up

In June 2016, the patient returned with a recurrent papule. The lesion had grown back 3 months before and was itchy upon touching. Histopathology of the biopsy showed a morphology nearly identical to that seen in the biopsy of 2014, now with slightly more intermixed eosinophils. The Ki-67 proliferation index reached up to 17.9%. The blood tryptase level was still low (4.55 µg/L). Due to the lack of complaints, a wait and watch strategy was proposed.

In August 2018, another punch biopsy was taken of the plaque due to irritation and slowly increasing size of the lesion (Figure 1B).

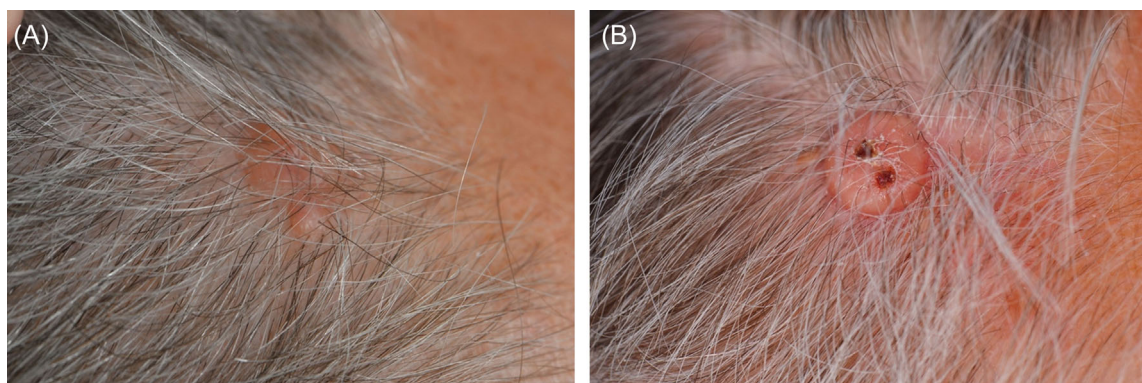


FIGURE 1 A, Clinical picture of the lesion in 2014, and B, the recurrent lesion in 2018

TABLE 1 Summary of the clinical course and laboratory, imaging, and histopathological characteristics

	2014	2016	2018
Physical examination			
Skin	Solitary plaque right temple. No other abnormalities	Recurrent solitary papule right temple. No other abnormalities	Regrowth to solitary plaque right temple. No other abnormalities
Therapy			
	Local excision	No (only punch biopsy)	Local reexcision
Laboratory			
Blood count	ND	ND	Normal
Renal function	ND	ND	Normal
Liver function	ND	ND	Normal
LDH (n < 248 U/L)	ND	ND	170 U/L
Serum tryptase (n < 11.4 µg/L)	2.9 µg/L	4.55 µg/L	4.37 µg/L
Bone marrow			
Flow cytometry	ND	ND	0.01% mast cells. CD2 and CD25 negative
Histopathology	ND	ND	No major or minor criteria for mastocytosis
Imaging^a			
CT scan thorax	ND	ND	No other localizations
MRI skull	ND	ND	No residual tumor
FDG-PET/CT scan	ND	ND	No metabolically active lesions
Molecular analysis			
NGS	No <i>KIT</i> mutation (skin lesion) ^b	ND	ND
Mutation specific PCR	ND	ND	No <i>KIT</i> D816V mutation (bone marrow)
Immunohistochemistry			
CD117	Positive	Positive	Positive
Tryptase	Positive	Positive	Positive
S100	Negative	Negative	Negative
MITF	Positive	Positive	Positive
CD25	Negative	ND	Negative
CD2	Negative	ND	Negative
CD30	Positive (heterogeneous)	Positive (heterogeneous)	Positive (heterogeneous)
Proliferation			
Mitotic figures	9/mm ²	ND	18/mm ²
Ki67	29.5%	17.9%	33.5%

Note: The major criterion for mastocytosis is the presence of multifocal clusters of mast cells (≥ 15 mast cells in aggregates) in bone marrow and/or other extracutaneous organs. Minor diagnostic criteria include elevated serum tryptase level, abnormal mast cell CD25 with or without CD2 expression, presence of *KIT* D816V mutation, and more than 25% of mast cells with atypical morphology in bone marrow or other extracutaneous organs.¹ Abbreviations: CT, computed tomography; FDG, fluorodeoxyglucose; LDH, lactate dehydrogenase; MITF, microphthalmia-associated transcription factor; MRI, magnetic resonance imaging; ND, not done; NGS, next generation sequencing; PCR, polymerase chain reaction; PET, positron emission tomography.

^aPerformed after reexcision of the tumor.

^bNGS included analysis of exons 8, 9, 11, 13, 14, and 17.

Histopathological examination showed a similar picture, still without epidermal involvement or a destructive growth pattern. However, there was a marked increase in mast cells with pleomorphic nuclei. Again, only a few spindle-shaped cells were found. Mitotic

activity was substantial, with up to 18 mitotic figures/mm², without atypical mitotic figures. In the Giemsa-stain, most neoplastic cells were hypogranulated. The Ki-67 proliferation index reached up to 33.5%.

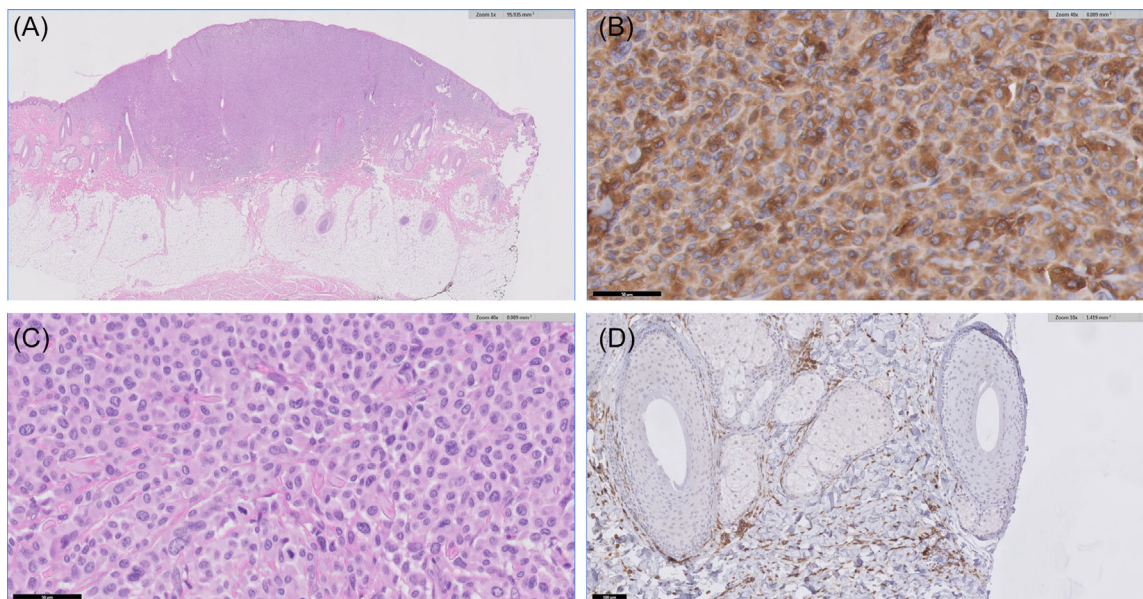


FIGURE 2 Skin excision in 2014. A, Overview of the lesion of diffuse sheets of epithelioid cells in the dermis (H&E, magnification $\times 10$). B, Positive tryptase staining (magnification $\times 400$). C, Details of the cells with some pleomorphism and scant mitotic figures (H&E, magnification $\times 400$). D, Small clusters of mast cells near the resection margin (CD117, magnification $\times 200$)

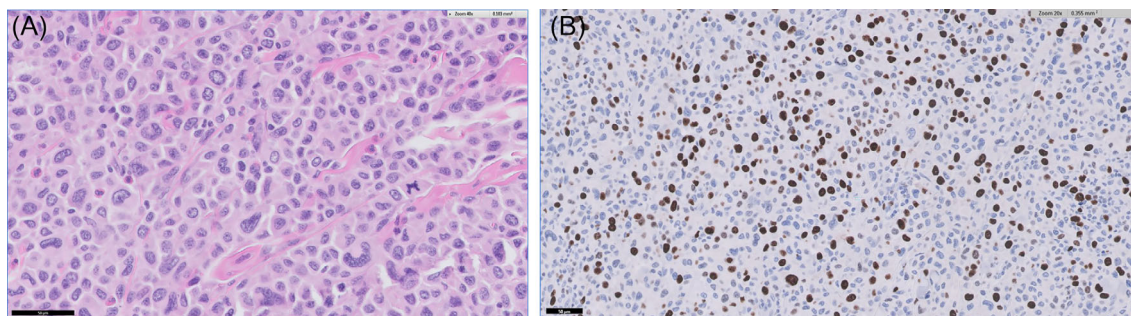


FIGURE 3 Skin excision of the recurrent lesion in 2018. A, Detail with progressive nuclear polymorphism and increasing mitotic rate (H&E, magnification $\times 400$). B, Focus with a high Ki-67 proliferation index (magnification $\times 200$)

In December 2018, reexcision with a wide local margin was performed. The histopathology was similar compared to previous studies, with a well demarcated lump containing predominantly pleomorphic mast cells (Figure 3A). Loose clusters of less atypical mast cells continued into the edges of the excision along the adnexa and vascular network, as in the previous excision. In a hot spot, 11 mitotic figures/mm² were found. The Ki-67 proliferation index was highly variable (<1% and in hot spots up to 25.3%) (Figure 3B).

The tryptase level remained low (4.37 $\mu\text{g/L}$). A bone marrow biopsy was performed in January 2019 and showed trilinear hematopoiesis with a slight increase in eosinophils. No mast cell aggregates or atypical mast cells were found. The bone marrow aspirate also did not show atypical mast cells, no co-expression of CD2 or CD25 on mast cells in flow cytometry, and no *KIT* D816V mutation. Postoperative magnetic resonance imaging of the skull did not show any residual mass. A computed tomography of the thorax did not show lung abnormalities or lymphadenopathy. Until October 2020 (last visit), no local recurrence or systemic involvement occurred.

3 | DISCUSSION

In the current case, atypical cutaneous mastocytoma and MCS are the main differential diagnostic considerations. The WHO classification defines cutaneous mastocytoma as “mostly pediatric lesions with sheets of mature looking mast cells ... without cytological atypia, which enables the distinction from an extremely rare MCS of the skin”; it defines MSC as “an extremely rare entity characterized by localized destructive growth of highly atypical mast cells.”^{1,22} In the registry of the European Competence Network on Mastocytosis, 2 out of 2985 adult patients (<0.1%) presented with MCS.²³ There are 13 cases of MCS or MCS-like neoplasms in adults reported in the English literature, including cutaneous and extracutaneous forms.^{24–33} Additionally, there are five reported cases of systemic mastocytosis with sarcomatous growth of mast cells, all in adults and extracutaneous.^{28,34–37} All MCS cases showed extensive local destructive growth, “metastases” and/or secondary systemic involvement, mostly in form of an MCL.^{24–38}

In our case, some features consistent with MCS were present, that is, recurrence of the lesion, (progressive) cytologic atypia, and pronounced mitotic activity. However, the lack of invasive growth, absence of secondary systemic involvement, and the long-term survival in our patient did not fit with the highly malignant behavior of MCS. On the other hand, recurrent lesions are rare in mastocytoma. Of the 14 cases in the literature with adult mastocytomas, eight had a surgical resection.^{4,5,7,10,11,13-15} In six of these eight cases, follow-up data were available, with durations between 7 months and several years, without reported recurrence.^{4,5,7,10,11,13-15} The recurrence in our case may be explained by small foci of mast cells in the resection margins. Although the WHO classification¹ states that atypia should be absent in mastocytoma, several case reports^{7,17-21} showed that type II atypia of mast cells can occur in mast cell tumors with benign behavior. The nuclear pleomorphism in our case seemed to be more pronounced than described in the previous case reports. A high mitotic activity was only described in one other case of a pleomorphic mastocytoma in an 11-year-old girl²¹ in whom no recurrence or progression occurred during 7 months of follow-up. Recently, a case of a pleomorphic mastocytoma was presented with distinctive genetic aberrations.³³ However, in contrast to our case, mitotic figures were not present and recurrence of the lesion was not mentioned. On the basis of the above-mentioned features, we argue that the lesion should be diagnosed as an atypical mastocytoma of the skin, borderline to a sarcoma. Because this is a mast cell tumor with unique features, the biological behavior cannot be predicted. We believe that close follow-up is warranted, which is further supported by three case reports.^{25,28,38}

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DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

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