CLINICAL REPORT

Acral myxoinflammatory fibroblastic sarcoma

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KEYWORDS
Sarcoma; Inflammatory; Acral; Myxoid; Bizarre cells

Summary  Acral myxoinflammatory fibroblastic sarcoma is a rare low-grade malignant soft tissue tumor, usually observed in the extremities of middle-aged patients. We report a case involving the third finger of the left hand of a middle-aged man. The tumor showed a nodular architecture, with cellular areas, occasional foci of hyalinized fibrosis, and hypocellular areas with a myxoid background. Various neoplastic cells were identified including spindled or rounded epithelioid cells and occasional bizarre giant cells, morphologically mimicking ganglion cells. Tumor cells were strongly immunoreactive for vimentin and variably positive for CD68 and CD34. The tumor was completely removed, without further treatment.

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Introduction

Acral myxoinflammatory fibroblastic sarcoma (AMFS) is a recently identified low-grade soft tissue tumor, mainly affecting the extremities of middle-aged subjects. We report a case in a middle-aged man, highlighting the diagnostic criteria for this rare tumor, which it is important not to overlook so as to be able to perform optimal surgical exeresis.

Clinical observation

A 51-year-old male, with no particular history, consulted for sensitive swelling of the inner side of the third finger of the left hand, which had been evolving for 6 months. Clinically, the lesion was nodular, soft, without local inflammatory signs or satellite adenopathy.

Coronal short T1 inversion recovery magnetic resonance imaging (STIR MRI) revealed isolated 3 cm tumefaction of the left third ray P1 soft tissue with normal bone medulla, with high contrast uptake, infiltration of surrounding tissue and erosion of the facing bone (Fig. 1). Biopsy exeresis enabled diagnosis, leading to amputation of the finger, ensuring the healthy status of the stump. Evolution was fair at 10 months’ follow-up.

Anatomopathology

The lesion was macroscopically nodular, without capsule, and of a yellowish white color (Fig. 2).

The microscopic aspect was lobular, poorly contoured and featuring often abrupt transition between dense cellular areas with an edematous or hyalinized background and hypocellular areas with a myxoid background. The tumor
cells were of medium to large size, and of two types: bizarre cells of a ganglion-like aspect (Fig. 3) and lipoblast-like vacuolated cells (Fig. 4). The mitosis index was low, at 3 per 50 high enlargement fields. A mixed inflammatory infiltrate of variable density associated lymphocytes, plasmocytes, and polymorphonuclear eosinophils and neutrophils (Fig. 5). Vessels were numerous outside the myxoid areas, with smooth and thick muscular walls. There was no necrosis.

Tumor cells were strongly immunoreactive for vimentin and variably positive for CD68 and CD34. The tumor was completely removed, without further treatment.

Immunohistochemistry showed strong and diffuse immunoreactivity for anti-vimentin antibodies and variable immunoreactivity for anti-CD68 and anti-CD34 antibodies. The tumor cells expressed no smooth muscle actin.

Discussion

AMFS was first described in 1998, almost simultaneously by three different teams, under three different names [1–3]. Some hundred cases have since been reported [3–7].
Acral myxoinflammatory fibroblastic sarcoma

It is a rare, low-grade sarcoma, mainly affecting middle-aged persons, and located in the extremities (fingers and hands, for 75% of cases). Clinically, the lesion is infiltrating, pain-free and of slow evolution.

Macroscopically, the tumor ranges in size from 1.5 to 18 cm, is generally well contoured, lobular, yellowish in color and sometimes mucoid. Occasionally, areas of necrosis and hemorrhage are found [8, 9].

Microscopically, there are three main characteristics [1, 3, 7, 10, 11]:

- a somewhat multinodular overall architecture, alternating densely cellular and myxoid hypocellular areas;
- mixed inflammatory infiltrate;
- bizarre giant and lipoblast-like cells.

The present case meets these microscopy criteria.

The immunohistochemical phenotype is non-specific: the tumor cells express vimentin (in all cases), CD68 (in 66% of cases), CD34 (in 28%), smooth muscle actin (in 6%) and cytokeratin (KL) (in 11%). They do not express epithelial membrane antigen, protein S100, desmin, CD15, HMB45, CD45 or Epstein-Barr virus (EBV) latent membrane protein (LMP1) [3, 7, 10].

The physiopathogenesis of AMFS is poorly understood. No infectious agent has been identified. PCR for EBV, performed in a few cases, indicated only latent rather than active infection [1, 3, 7]. Immunohistochemistry with anti-cytomegalovirus (CMV) and/or anti-EBV antibodies, performed in several series, also proved negative [1, 3, 8, 11]. A cytogenetic study of a single case found a complex karyotype with reciprocal t(1;10)(p22, q24) translocation, and three and 13 deletion [3, 6].

From the clinician’s point of view, differential diagnosis is between infectious or chronic inflammatory process and benign tumor such as nodular fasciitis, tendon giant cell tumor and synovial pseudocyst. From the pathologist’s point of view, differential diagnosis depends on the predominance of inflammatory, myxoid and bizarre cell components, and is between non-tumoral lesions such as inflammatory pseudo-tumor (bizarre giant cells being sometimes difficult to identify in the inflammatory infiltrate) and tumoral lesions such as giant cell tumor, inflammatory fibrosarcoma, inflammatory myofibroblastic tumor or myxofibrosarcoma [1, 3, 7, 8, 10, 11]. Myxofibrosarcoma differs from AMFS in its proximal location and it is more aggressive, pleomorphic and storiform character [4, 7]. Alternative diagnoses can often be ruled out by the discovery of bizarre giant cells and by the tumor location [1, 7]. In practice, sarcoma of the extremities is rare, and mainly limited to epithelioid sarcoma, synovial sarcoma and clear-cell sarcoma [1, 7, 12].

Local recurrence ranges from 22% [1, 3, 7] to 67% [3, 7, 10], at a median of 4 to 10 months, depending on the series. Only three cases of metastasis have been reported: two ganglionic [3, 4, 10] and one pulmonary [3, 10], the latter not demonstrated on histology. No associated deaths have been reported.

Treatment consists in full surgical exeresis, checking the healthy status of the remaining edges [9].

Conclusion

AMFS is a rare low-grade tumor associated with virtually no metastasis, but liable to recurrence. Prognosis depends on initial surgical management, which should consist in complete exeresis, despite the probable functional impact.

Several clinical issues remain unresolved, especially regarding recurrence and metastasis rates and intervals. Further research on these lines is underway to assess results more clearly.

Conflicts of interest statement

None.

References