Intermediate-Grade Myxofibrosarcoma Mimicking a Blistering Disease

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To the Editor

Malignant fibrous histiocytoma is one of the most common sarcomas of soft tissue in old age and groups together a series of histologic variants—pleomorphic (60%), myxoid (25%), angiomatoid, giant-cell, and inflammatory. Some authors consider myxofibrosarcoma to be a myxoid variant of malignant fibrous histiocytoma, whereas others maintain that it is an independent entity.

Malignant fibrous histiocytoma typically manifests as a soft and painless nodular mass in the subcutaneous tissue of the lower limbs in elderly patients. Other clinical variants have been reported, such as diffuse infiltration, multiple nodules, or papules, and even 1 case that mimicked papulosis. The tumor originates in the subcutaneous tissue, often extends to deeper layers (90%); extension to upper layers (dermis) is uncommon, with fewer than 10 cases reported in the literature. A series of histologic grades has been reported, and variants with a sparse myxoid matrix and greater cellularity, pleomorphism, and number of mitoses have been considered high-grade. Lower-grade variants generally have an abundant myxoid matrix in the superficial layers, and this necessitates deep incisional biopsies to reach layers.

Something to think about: If Dr Betlloch’s impression is confirmed, the last link in the chain, that of health care provision, might successfully implement—the latest buzzword—the objectives to which the public health system allocates many costly mechanisms, players, and resources and which it fails to achieve time and time again, to reduce the waiting list and create a more appropriate distribution of the time dedicated to patient care. This is yet another demonstration of the enormous potential—often underestimated—of dermatologists to make an impact.

Common sense tells us that, if we can improve our work situation, then this can only be to the advantage of our patients.

References

with atypical cellularity and enable correct diagnosis. Differential histologic diagnosis is with stasis dermatitis, superficial angiomyxoma, mucinosis, and generally with conditions involving mucin aggregation.

The treatment of choice for malignant fibrous histiocytoma involves wide excision, and radiotherapy has only proven useful to reduce the risk of local recurrence. Five-year survival is almost 85% in less aggressive variants of this tumor. We present the case of a 90-year-old man with a history of chronic venous insufficiency and essential thrombocythemia under treatment with hydroxyurea. He presented with a 1-year history of tumefaction in the left leg that varied throughout the day and that later became persistent as it progressed towards the foot. Translucent, blister-like lesions gradually developed on this swollen area and were accompanied by local pain.

The most remarkable finding in the physical examination was the presence of varicose veins and acroangiodermatitis, supporting the previous diagnosis of chronic venous insufficiency; swelling was also noticed on the left leg (Figure 1). A translucent papulous aggregate, which was gelatinous on incision, was observed on the medial aspect of the left leg (Figure 2).

Standard laboratory tests were requested: complete blood count, biochemistry, iron profile, and prostate-specific antigen. These revealed only macrocytosis (mean corpuscular volume, 107.9 fl) and thrombocytosis ($571 \times 10^3/\mu L$), which was attributed to the essential thrombocythemia and treatment. Doppler ultrasound ruled out venous obstruction and magnetic resonance imaging showed diffuse infiltration of the subcutaneous tissue that was compatible with an infiltrating lesion.

A first biopsy revealed separation of collagen bundles and dilation of superficial and deep vascular plexuses. Localized chronic lymphedema was diagnosed.

This inconsistency between clinical and histologic data led to a second biopsy, which revealed a massive aggregate with a slightly basophilic matrix causing excessive elevation of the epidermis/dermis that shaped the translucent papules observed on examination. The myxoid matrix extended from the subcutis almost to the papillary dermis. In addition, cells with a pleomorphic appearance and mitotic figures were observed (Figure 3)—these increased in number in deeper layers. A cytoplasmic mucin aggregate was also observed. Immunohistochemical analysis revealed positivity for vimentin and CD34.

These data indicated intermediate-grade malignant fibrous histiocytoma with extension to the dermis. The analysis of tumor extension (computed tomography of the thorax, abdomen, and pelvis) did not reveal metastasis to the lymph nodes or solid organs, and the tumor was staged as localized.

Given the patient’s age and the localized nature of the disease, local radiotherapy was administered up to a total dose of 3000 cGy in 10 fractions. Tolerance was good and there was a visible partial response after the first sessions, with disappearance of the
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blister-like lesions and reduction of the infiltration. This benefit, however, was merely transitory (3 months), and subsequent progression necessitated supracondylar amputation. Six months after surgery, there were no signs of the disease.

We present a case of malignant fibrous histiocytoma with a peculiar clinical presentation in the form of aggregated translucent papules containing mucin. The clinical condition mimicked a blistering disease, although the possibility of a lymphatic neoplasm or a disease involving deposition of amorphous acellular material such as mucin was also considered. We were unable to find cases with a similar presentation in the literature. Another peculiarity of our case is the extensive dermal involvement at onset, a finding which is relatively uncommon.2,3,6 Furthermore, as occurred with our case, errors in the initial histologic diagnosis are common. Therefore, a higher level of suspicion and deep incisional biopsy are necessary to rule out this tumor2 and to carry out a differential diagnosis with other entities that display mucin aggregation or similar histologic patterns (chronic lymphedema or stasis dermatitis).

References

Study of Urgent Referrals to the Dermatology Department of a Referral Hospital

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To the Editor:

Dermatology is not a specialty in which urgent or priority consultations make up a large part of routine practice. However, whether due to long waiting lists or to the poor level of health education among the population, dermatologists are treating an increasing number of patients who have been referred as priority cases. We therefore undertook a study to analyze the priority referrals to our department as an aid to planning and improving the quality of care provided.

Data were collected prospectively in 1998 on the priority patients who had been referred by primary care clinics to dermatology clinics associated with Hospital Miguel Servet in Saragossa, Spain. The population of the health care area is 533,946 inhabitants, consisting of an urban population of 474,523 and a rural population of 59,019.

A record was designed for each patient using the Access database application. The records included the patient's identification number, date of birth, and sex, whether the patient was from a rural or urban area, whether the patient was classified as ordinary or priority, the date of consultation, and the diagnosis. A database was created that contained the possible diagnoses and their codes according to the International Classification of Diseases, Tenth Revision.1 Statistical analysis of the data was performed using the SPSS statistical software package and the Excel spreadsheet. The relationship between qualitative variables was analyzed using the #c2 test and the