Solitary fibrous hemangiopericytoma of atypical location: importance of immunohistochemical study

ABSTRACT

Background: The rare cutaneous solitary fibrous tumor was initially described in the thoracic cavity in relation to the pleura and subsequently has been associated with other serous membranes. It has been described in other extraserosal locations including the skin. Knowledge of its existence along with fairly typical histological features and the immunohistochemical expression pattern with intense positivity for CD34 allow the increasing diagnosis of this condition, which suggests that these cases were not previously diagnosed as such.

Clinical case: We report the case of a 43-year-old male with a painless nodule in the first left finger pad clinically suggestive of pyogenic granuloma or nodular melanoma, which was diagnosed by excisional biopsy and immunohistochemical study as a solitary fibrous tumor.

Discussion: Only 11 cases of cutaneous solitary fibrous tumor have been published in the following locations: head, cheek, thigh, chest, back and nose. Our work describes the first case of cutaneous solitary fibrous tumor in the hand. Solitary fibrous tumor derived from mesenchymal cells expresses CD34 and hence its presentation in any location. In our case it was in the hand. It explains the problems encountered in the clinical differential diagnosis with other tumors such as nodular melanoma, pyogenic granuloma, giant cell tumor of tendon sheath, fibroma, benign peripheral nerve sheath tumors, etc. As we consider the histology, differential diagnosis should be made with other tumors that also express CD34.

Conclusions: Solitary fibrous tumors derived from mesenchymal cells express CD34 and hence its presentation in any location. In our case it was in the finger pad.

Key words: Cutaneous solitary fibrous tumor, CD34, fibrous tissue neoplasms.
ABSTRACT

Solitary fibrous tumor is a rare spindle cell neoplasm, initially described in the pleura, although it was subsequently found in other locations. Few cases of solitary fibrous tumor on the skin have been published. It is possible that it is an underdiagnosed disease in this location given its clinical and histological similarity with other tumors. Greater knowledge of its existence, together with a very suggestive histological image and typical immunohistochemical expression, allow for them to be diagnosed in new and multiple locations. We report a case of patient with a solitary fibrous tumor on the ball of the finger and the clinical and histological differential diagnosis is discussed.

CLINICAL CASE

A 43-year-old male without significant personal or family history presented to the Dermatology Service due to a painless, erythematous nodule on the ball of the first finger of the left hand not attached to deep planes (Figures 1A-1C). The patient reported that the lesion appeared 3 years prior as a small, erythematous lesion that grew slowly and progressively. On examination he appeared to be in good general health without weight loss or local adenopathy. Blood tests found no alterations (glucose 102 mg/dL). Ultrasound demonstrated a well-defined 25 × 15-mm nodule, which resembled an angioma. On skin examination a lesion of non-melanocytic appearance without a defined pattern was seen (Figure 1D). Under local anesthesia, excision of the lesion was carried out and the defect was closed. The macroscopic finding was that of a well-defined, whitish nodular specimen of 1.5 × 1 cm and covered by whitish-yellow skin on the entire surface. The nodule was of elastic, gelatinous consistency with a smooth surface. Histological analysis of the specimen showed a well-defined, nonencapsulated deep skin tumor with grouping of spindle-shaped cells, without atypia and with thick collagen bundles interspersed in the stroma and abundant small and dilated vessels (Figures 2A and 2B). The overlying epidermis showed no significant alterations. Immuno-histochemical study showed positivity for vimentin, CD34, caldesmin and weakly for BCL-2, as well as negativity for PS100 and desmin (Figure 2C). Diagnosis of solitary fibrous tumor was made with all these findings. The patient whose case we describe here did not present recurrence or relapse of the disease after 1-year follow-up.

DISCUSSION

Solitary fibrous tumor is a rare spindle cell tumor described for the first time in the visceral pleura. Since 2003, cases of extrapleural location have been published and due to its origin in undifferentiated mesenchymal cells with morphology similar to a fibroblast, it can be found on the skin. Its cells express CD34 and participate in the antigenic presentation in the connective tissue.
With respect to its location, there have been 11 cases published in the skin: six in the head and neck, one in the cheek, thigh, chest, back and nasal cavity. This is the first case reported of a hemangiopericytoma solitary fibrous tumor in a finger of the hand.

Knowledge of its existence together with the characteristic histological report and the immunohistochemical pattern of expression allow for this tumor to be described in multiple and new locations.

The tumor tends to be histologically well defined with smooth margins and surrounded by a normal thin epidermis as a pseudoencapsulation. It is composed of spindle cells and thick collagen bundles interspersed in the stroma. Vascularization is prominent; therefore, it is not difficult to confuse it with a tumor of vascular origin.

It is necessary to establish the differential diagnosis with other spindle-cell skin neoplasms that express CD34 such as dermatofibrosarcoma protuberans, neurofibroma, schwannoma, solitary fibrous tumor/hemangiopericytoma, epithelial sarcoma, sclerotic fibroma, spindle cell lipoma, low-grade fibrosarcoma, etc.

It was especially relevant in this case to make the clinical differential diagnosis with nodular melanoma, pyogenic granuloma or pigmented basal cell carcinoma. As a result of immunohistochemical study there is usually no difficulty in recognizing solitary fibrous tumor and differentiating it from other lesions using CD34 positivity (which is considered an absolute for diagnosis) and BCL-2, as well as negativity for other specific markers such as PS100. Therefore, the CD34 marker is very sensitive for detecting solitary fibrous tumor, but with low specificity. In recent investigations it has been demonstrated that the BCL-2 oncoprotein, one of the many molecules that regulates apoptosis, is strongly expressed in the solitary fibrous tumor, whereas dermatofibroma and smooth muscle tumors are negative for this oncoprotein. In other tumors such as the dermatofibroma protuberans and the neurofibroma, BCL-2 expression is variable; therefore, the morphology is of great importance and the immunohistochemical expression of the BCL-2 protein can be used to confirm, together

Figure 2. (A) Well-defined, nonencapsulated tumor located on the deep dermis (panoramic view x5, H&E). (B) Solitary fibrous tumor is comprised of a grouping of many small dilated spindle cells with thick collagen bundles interspersed in the stroma (x15, H&E). (C) CD34 positivity is very specific for this tumor (x15, CD34).
with the positivity of CD34, diagnosis of solitary fibrous tumor. The cells of this tumor also stain with CD99, which helps to establish the differential diagnosis.\(^5\)

Use of immunohistochemical markers is fundamental for the exact classification of these tumors. Solitary fibrous tumor is not usually immunoreactive for S-100, keratin, actin, desmin, factor VIII antigen, CD68, type IV collagen and epithelial membrane antigen. In recent immunohistochemical and ultrastructural studies it has been demonstrated that these tumors derive from undifferentiated mesenchymal cells, without mesothelial CD34+ differentiation, known as interstitial dendrocytes that participate in the antigenic presentation in the connective tissue.\(^7,\,9\) This would explain that the tumor might grow in any area of the body, including the skin.

In the case studied, according to the PS100 marker being negative, diseases such as neurinoma, schwannoma, spindle-cell lipoma and melanoma were able to be ruled out. The desmin negativity allows ruling out muscular tumors such as dermatofibrosarcoma, which is a non-muscular fibrohistiocytic tumor.

The solitary fibrous tumor tends to follow a benign course and, in fact, various authors do not assign it a malignant behavior;\(^3,\,7\) however, in other series, clear examples of malignant behavior have been recognized (estimated incidence >10%) that are attributed to a short follow-up period, failure in the diagnosis of malignant cases or to shorter series.\(^9,\,10\) Malignant cases have been found through the sarcomatous differentiation such as liposarcoma.\(^11\) Either way, there should be a typical component of solitary fibrous tumor so that it can be considered malignant.\(^5\) In these cases an increase in the expression of p53,\(^12\) Ki67 and CD31 has been found.\(^11\) Malignant transformation appears to be associated with a differentiation revealed by the loss of CD34 expression. Of the solitary skin fibrous tumors reported, none has shown malignant behavior. In this patient, Ki67 and CD31 tests were done and were negative. The prognosis is unpredictable because the tumor may recur or metastasize, even when it lacks malignant characteristics. For this reason, wide surgical excision is recommended and long-term clinical and radiological follow-up in all cases, whether benign or malignant. Some authors claim that it is not wise to consider it as definitely benign;\(^5,\,9\) however, it is not recommended that co-adjuvant treatment be administered. Surveillance only is recommended such as in the case presented, which at 1 year of follow-up has not demonstrated recurrence or relapse.

In conclusion, this is a little-known tumor that makes the differential diagnosis with other more common tumors necessary because of its spindle-cell characteristics and for which immunohistochemical study is of vital importance. Although the majority of solitary fibrous tumors are reported as benign and no malignant cases of the skin have been described, the prognosis is not known with any certainty, a reason for which long-term follow-up is necessary.

REFERENCES


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