
Inflammatory Pseudotumor of the Skin: A Case Report And Review Of The Literature

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Citation

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Abstract

Inflammatory pseudotumor is a rare mesenchymal neoplasm most commonly developing in the viscera with very rare primary cutaneous involvement. This neoplasm, though usually behaving in a benign fashion, is actually considered to have intermediate biologic potential, as cases of local recurrence and death have been reported. We report the case of a 70-year-old male with a cutaneous inflammatory pseudotumor presenting on the thigh, which after biopsy and histologic identification, was completely excised. A review of this entity and other cases of inflammatory pseudotumor of the skin is presented.

INTRODUCTION

Though inflammatory pseudotumor (IPT) was first recognized in the lung, it has since been identified in many other visceral organs; although primary cutaneous involvement is rare. (1,2) Inflammatory pseudotumor was the name first used to describe this reactive inflammatory lesion that resembles a neoplasm clinically and histologically. (3) Currently, the term IPT designates lesions that have a spindle cell proliferation in association with a mixed inflammatory infiltrate. (3) Some of the cases are reported as inflammatory myofibroblastic tumors; there has been much discussion regarding the terminology that should be used for these neoplasms.

IPT has considerable heterogeneity in its microscopic features. (4) Its components, namely spindle cells, a mixed inflammatory infiltrate, and collagen bundles, can exist in varying proportions in different patients and sometimes within the same lesion. (5) A study by Coffin et al, classified 3 distinct patterns: the myxoid or vascular pattern, the compact spindle cell pattern, and the hypocellular fibrous pattern. (5) The specific classification for each lesion is determined by the amount of each component it contains. (3,6)

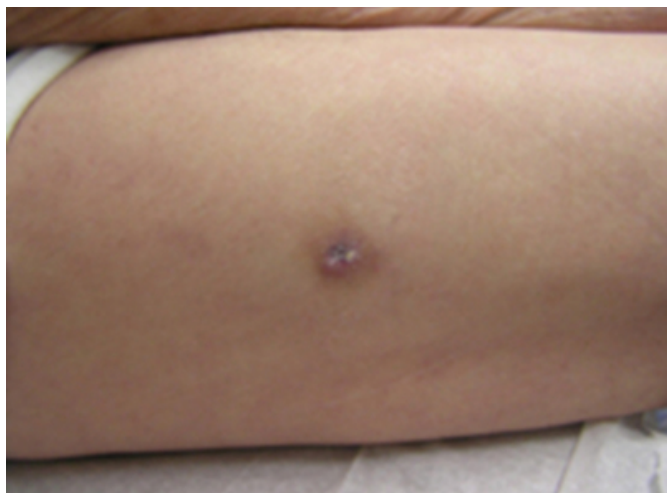
Although IPT has a favorable prognosis, cases of malignant transformation, local recurrence, and transformation to an undifferentiated sarcomatoid proliferation have been reported, so it is considered to have intermediate potential. (2,7,8) However, these complications have not been reported

in cases of cutaneous IPT. In most cases, the standard therapeutic approach is complete surgical excision, but in more complicated cases, it may be necessary to consider adding adjuvant therapies such as chemotherapy or radiation. (8) We describe a case of IPT in the skin of a 70-year-old male that was treated surgically with good outcome.

CASE REPORT

A 70-year-old man presented with a 3-month history of an asymptomatic, but growing nodule on his right thigh. The patient was healthy and had no complaints of fever, weight loss, or night sweats. He could not recall any preceding injury in the area. Physical examination revealed a 1.5 cm red-brown, minimally elevated, firm nodule with some overlying scaling (Figure 1). The primary clinical differential diagnosis considered included a giant dermatofibroma versus dermatofibrosarcoma protuberans.

Figure 1



Initial investigation included a lesional punch biopsy which revealed spindle cells in a fascicular pattern with a mixed lymphoplasmacytoid inflammatory infiltrate and thickened collagen bundles (Figure 2, 3, 4).

Figure 2

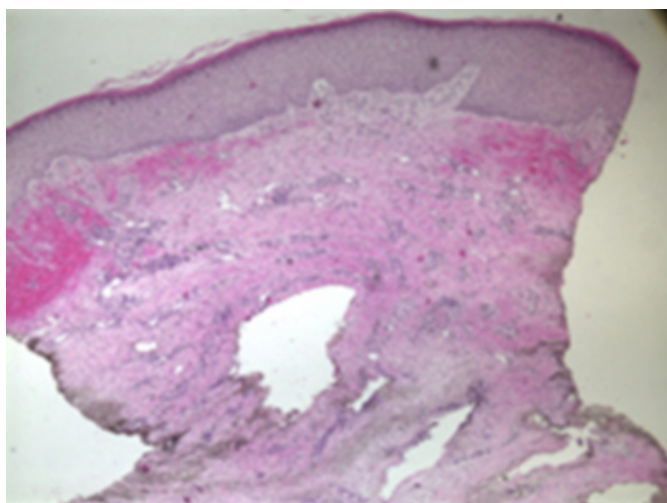


Figure 3

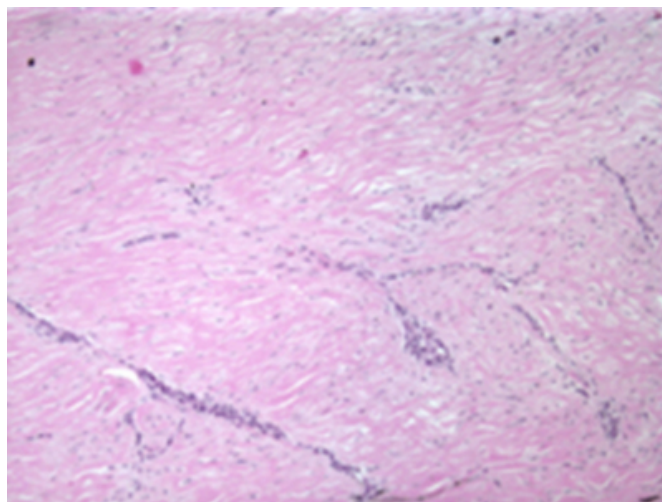
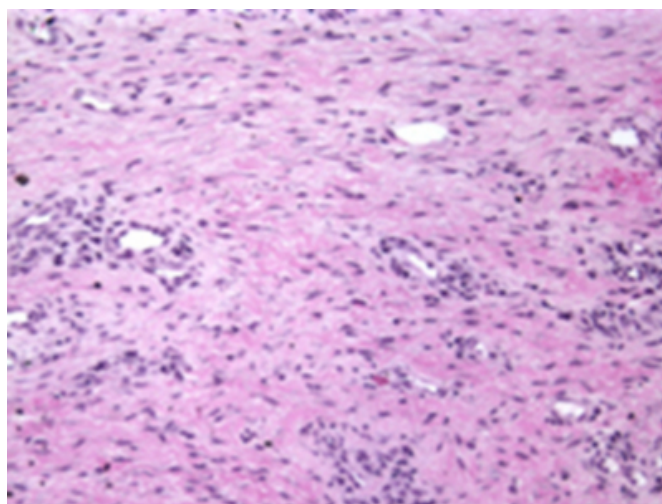


Figure 4



These histologic findings led to a diagnosis of IPT. Elliptical excision of the nodule with 5 mm safety margins was performed. By 3 months postoperatively, no recurrence was evident.

DISCUSSION

Inflammatory pseudotumor is a rare, intermediate behavior, mesenchymal neoplasm that is uncommon in the skin. (1,2,8) From a review of the English language literature, we identified 11 additional reports of cutaneous IPT (Table I).

Figure 5

Table 1: Clinical Features of Cutaneous IPT*

| Study | Patient No. | Age(yr)/ sex | Original Classification | Location | Size (cm) | Treatment | Outcome |
|--|-------------|--------------|-------------------------|----------|-----------|-----------|-----------------------|
| Hurt et al., 1990 ¹⁰ | 1 | 19/F | IPT | Neck | 1.4 | Excision | No recurrence, 5 yr. |
| | 2 | 39/M | IPT | Arm | 0.8 | Excision | No recurrence, 5 yr. |
| | 3 | 40/F | IPT | Calf | 0.9 | Excision | No recurrence, 4 yr. |
| | 4 | 64/M | IPT | Arm | 1.3 | Excision | No recurrence, 2 yr. |
| Vadmal et al., 1999 ⁷ | 5 | 49/F | IMT | Elbow | 4.0 | Excision | No recurrence, 6 wks. |
| Yang et al., 1993 ¹¹ | 6 | 44/M | IPT | Hand | 4.0 | Excision | No recurrence, 8 mo. |
| El-Shabrawi-Caelen et al., 2004 ⁶ | 7 | 56/F | PCG | Wrist | 1.0 | Excision | No recurrence, 73mo. |
| | 8 | 56/F | PCG | Arm | 2.0 | Excision | No recurrence, 33mo. |
| Present Case | 9 | 15/M | PCG | Shoulder | 1.0 | Excision | No recurrence, 13mo. |
| | 10 | 89/M | IMT | Shoulder | 5.0 | Excision | No recurrence, 16mo. |
| | 11 | 70/F | IMT | Scalp | 2.0 | Excision | No recurrence, 54mo. |
| Present Case | 12 | 70/M | IPT | Thigh | 1.5 | Excision | No recurrence, 3 mo. |

* F indicates female; M, male; IPT, inflammatory pseudotumor; PCG, plasma cell granuloma; IMT, inflammatory myofibroblastic tumor

Although visceral IPT occurs more frequently in children, previously reported cases of primary cutaneous IPT occurred primarily in adults. (1,4,9,10) The ages of the patients identified in our review ranged from 15 to 89 years (mean, 51 years). (1,4,9,10) The male to female ratio in these cases is equivalent at 1:1. (1,4,9,10) The lesions were solitary and occurred most commonly on the extremities. (1,4,9,10) In all of the cases, treatment included surgical excision, with no cases of recurrence reported. (1,4,9,10)

There is still ongoing investigation into the etiopathogenesis of IPT. In many studies, the term inflammatory pseudotumor is used to describe a variety of lesions. (4) However, a recent report by Shabrawi-Caelen et al, concluded that cutaneous IPT is a likely a broad category that encompasses plasma cell granulomas and inflammatory myofibroblastic tumors. (4) This study reviewed 5 cases of cutaneous IPTs. (4) They found that the lesions fit 2 histologic patterns. (4) The first had a dense inflammatory infiltrate with plasma cells, but no myofibroblasts were identified. (4) The second pattern had spindle cells which were identified as myofibroblasts with a similar inflammatory infiltrate. (4) The authors concluded that the first pattern, without identified myofibroblasts, is better termed plasma cell granuloma (PCG). (4) The second pattern is best referred to as inflammatory myofibroblastic tumor (IMT) – the term IPT encompasses both of these patterns. (4)

The exact etiopathogenesis has still not been defined for IPT after intense investigation. (6,7) Historically, there was disagreement in regards to whether IPT represents a true neoplasm or an aberrant inflammatory response to an

infectious or noninfectious agent. (6,7) Infections may be associated with the development of one group of IPTs, which appears to be reactive in nature. (6) Infectious agents may be certain viruses and bacteria, including: human herpesvirus-8, Epstein-Barr virus, Pseudomonas species, and mycoplasma. (6) The majority of the IPT likely belong to the group that is considered to have a neoplastic pathogenesis because of identified genetic abnormalities and potential for aggressive behavior. (6) Some IMTs have chromosomal abnormalities, including fusion of the anaplastic lymphoma kinase (ALK) gene to tropomyosin 3 or tropomyosin 4. (11,12) Moreover, a transcript involving ALK and the clathrin heavy chain genes was described in the pathogenesis of some IMTs. (12) DNA ploidy analysis has been performed in some studies; it was initially done as a marker of neoplasia, but may also be useful to determine prognosis. (2,13) In one study by Biselli et al, 5 of 9 tumors were diploid, 4 were hyperdiploid, and in about half of the cases, aneuploidy was associated with a more aggressive tumor behavior. (13) These studies were all done in the IMT subtype of IPT.

Although IPT is generally considered as benign or having low malignant potential, local recurrence has been reported in 25 to 37% of cases. (12) Recurrence is observed most frequently in cases with primary intraabdominal involvement; it has also been demonstrated that with each recurrence, the tumor may become more poorly differentiated. (12) There have also been recent reports of malignant transformation and of malignancy arising in the residual tumor. (7)

In light of these possible adverse outcomes, most IPT are treated with complete surgical excision. (1,4,8,9) When IPT is located in visceral organs, complete excision may be anatomically impossible. In these cases, other adjunctive treatments including chemotherapy or radiation may be used. (14) Even nonsteroidal anti-inflammatory medications have been shown to help reduce the size of IPT by interfering with angiogenesis. (14) Most cutaneous IPT can be completely resected, so other more aggressive treatments have not been necessary to date.

In all of the cutaneous cases of IPT reported, surgical excision has been the course of therapy and no complications or recurrences were reported. At this time, complete surgical excision seems to be the best approach to management of primary cutaneous IPT. Further study may, in time, better define the nature of this neoplasm and the optimal treatment approach.

ABBREVIATIONS

IPT: Inflammatory pseudotumor
IMT: Inflammatory myofibroblastic tumor
ALK: Anaplastic lymphoma kinase
PCG: Plasma cell granuloma

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