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A Review for Dermatofibrosarcoma Protuberans

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) stands as the most prevalent cutaneous sarcoma, marked by slow growth and a protuberant papulonodular appearance. Although rare, its impact is significant, necessitating a comprehensive understanding of its characteristics and optimal management. DFSP typically affects adults, though age variability exists, presenting diagnostic challenges. Histological evaluation, often utilizing CD34 immunohistochemical staining, is crucial for accurate diagnosis. The primary treatment modality remains complete surgical excision, emphasizing the importance of generous margins. Imatinib emerges as a valuable therapeutic option for unresectable or metastatic cases. Complementary approaches such as adjuvant radiation therapy and Mohs surgery enhance the treatment armamentarium, allowing for a tailored, multidisciplinary strategy. Regular follow-up is essential for monitoring and addressing potential complications. While local recurrence is a concern, DFSP generally carries a favorable prognosis, especially with early detection and appropriate intervention. Ongoing research continues to refine treatment paradigms, promising improved outcomes for individuals affected by this distinctive cutaneous sarcoma.

KEYWORDS: Dermatofibrosarcoma protuberans, cutaneous sarcoma, CD34, diagnosis, treatment, imatinib, surgical excision, adjuvant radiation therapy, Mohs surgery, multidisciplinary approach, recurrence, prognosis.

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INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) represents a distinct type of soft tissue sarcoma that is characterized by its low to intermediate grade [1-3]. The tumor typically exerts its effects through local extension into nearby structures. Notably, a subset of DFSP cases (about 5% to 10%) may exhibit a fibrosarcomatous component, indicating a more aggressive nature. Furthermore, approximately 5% of all DFSP cases may lead to metastatic disease, with the lungs and lymph nodes being the most commonly affected sites [4, 5].

Dermatofibrosarcoma protuberans (DFSP) stands out as the most prevalent cutaneous sarcoma, despite being a rare neoplasm in the broader context. Its estimated incidence is around 4.2 cases per million persons per year [1-3, 6]. The precise etiology of this neoplasm remains a subject of debate, but there is a likelihood that it originates from CD34+ dendrocytes, which are typically present in the dermis [7]. The majority of DFSP cases exhibit a reciprocal translocation between chromosomes 17 and 22, or, less commonly, a supernumerary ring chromosome formed from segments of chromosomes 17 and 22 [8, 9].

While DFSP is most commonly diagnosed in individuals aged 20 to 59, it can manifest at birth or occur in older adults, highlighting its variable age distribution. The unique characteristics and incidence patterns of DFSP underscore the need for further research and understanding of this intriguing cutaneous sarcoma [10, 11].

DFSP tends to occur most frequently on the trunk but can also be found on the arms, legs, or head and neck. The diagnostic process involves the performance of a large, deep punch biopsy (6 or 8 mm) or an incisional/excisional biopsy. Importantly, a shave biopsy is not recommended when DFSP is being seriously considered for diagnosis due to the risk of inadequate sampling [12, 13].

Treatment for DFSP primarily revolves around complete surgical removal of the tumor, with recommended margins ranging from 2 to 4 cm. This surgical approach is widely adopted as the standard of care to ensure comprehensive excision and optimize outcomes for individuals diagnosed with DFSP [14, 15].

DIAGNOSIS

Dermatofibrosarcoma protuberans (DFSP) predominantly occurs on the trunk, although it can also manifest on the arms, legs, or head and neck. Characteristically, DFSP presents as a gradual, exophytic papule that progressively enlarges, giving rise to additional papules and nodules of varying sizes that protrude above the skin surface. Notably, atrophic plaque-like forms, devoid of a protuberant appearance, are not uncommon [12, 13].

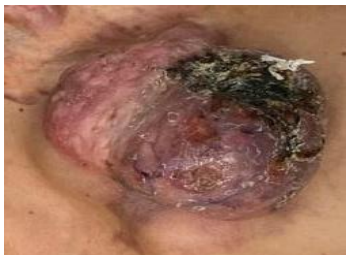

Figure 1. Chest area DFSP

DFSP lesions typically exhibit a skin-colored hue, but variations include red, pink, or white tones, particularly in atrophic areas. While DFSP is generally asymptomatic, a subset of patients, ranging from 10% to 20%, may experience local pain or discomfort associated with the lesion [16].

An uncommon variant of DFSP known as Bednar tumor may display pigmentation due to the presence of admixed melanocytes. These diverse clinical presentations underscore the importance of recognizing the varied features of DFSP for accurate diagnosis and appropriate management [5].

The diagnosis of dermatofibrosarcoma protuberans (DFSP) is typically confirmed through a comprehensive histological examination, often involving a large, deep punch biopsy (6 or 8 mm) or, in some cases, an incisional or excisional biopsy. It is crucial to note that a shave biopsy is not recommended when DFSP is being seriously considered diagnostically [12, 13].

Immunohistochemical staining plays a pivotal role in the histologic evaluation of DFSP. Nearly all DFSPs exhibit positive staining with CD34, a specific marker, contributing to the diagnostic process. This immunohistochemical characteristic aids in accurately identifying and confirming DFSP, facilitating appropriate therapeutic decisions [17].

The differential diagnosis of dermatofibrosarcoma protuberans (DFSP) encompasses various skin conditions and soft-tissue tumors, including dermatofibroma, hypertrophic scar, keloid, and other entities. The suspicion of DFSP often arises based on the clinical history of a slow-growing tumor with a distinctive protuberant papulonodular quality. Clinicians consider these differential diagnoses to distinguish DFSP from other benign or malignant skin lesions, emphasizing the importance of a comprehensive evaluation for an accurate diagnosis and subsequent management [18].

TREATMENT

In cases of dermatofibrosarcoma protuberans (DFSP) where tumors are unresectable or have metastasized, imatinib stands out as the preferred drug therapy. Imatinib, a tyrosine kinase inhibitor, has demonstrated efficacy in managing advanced or difficult-to-operate cases of DFSP [5].

The primary treatment procedure for DFSP involves complete surgical removal of the tumor, ensuring margins of 2 to 4 centimeters. This surgical approach is the most commonly employed and has proven effective in addressing localized forms of the disease [14, 15].

In certain situations, particularly for specific cases, adjuvant radiation therapy may be recommended. This additional treatment modality is considered based on individual patient factors and the characteristics of the tumor [19].

Mohs surgery, known for its precision in excising skin cancers layer by layer, has also been utilized in treating DFSP, showing some degree of success. Mohs surgery may be considered in specific instances where its advantages align with the patient's condition [20].

The choice of treatment, whether surgical, pharmaceutical, or a combination, is tailored to the specific circumstances of each patient, emphasizing a multidisciplinary approach that involves dermatologists, surgeons, oncologists, and other specialists. This collaborative effort aims to optimize therapeutic outcomes, minimize recurrence, and manage potential complications associated with DFSP. Regular follow-up is integral to monitoring the patient's progress and adjusting the treatment plan as needed [5, 14, 15].

CONCLUSION

dermatofibrosarcoma protuberans (DFSP) poses a unique challenge in the realm of cutaneous malignancies, being the most common cutaneous sarcoma despite its overall rarity. Characterized by slow growth and a protuberant papulonodular appearance, DFSP primarily affects adults but can present at any age. Diagnosis relies on careful histological evaluation, often marked by positive CD34 immunohistochemical staining.

The cornerstone of treatment remains complete surgical excision with generous margins, showcasing its efficacy in managing localized disease. For cases where surgical resection is challenging or in the presence of metastatic disease, imatinib emerges as a key therapeutic option, demonstrating notable success.

Adjuvant radiation therapy and Mohs surgery may complement the treatment arsenal, highlighting the importance of a tailored, multidisciplinary approach. Regular follow-up is crucial for monitoring disease progression, ensuring optimal outcomes, and addressing potential complications.

Despite its potential for local recurrence, DFSP generally carries a favorable prognosis, especially with early diagnosis and appropriate intervention. Ongoing research and clinical advancements contribute to refining treatment strategies, offering hope for improved outcomes in the management of this unique cutaneous sarcoma.

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