ISSN : 2347 - 2243

Indo - American Journal of Life Sciences and Biotechnology

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ISSN 2347-2243 www.iajlb.com

Indo-Am. J. of Life Sc & Bt.2023 Vol.20, Issue3, 2023

Indo-American Journal of Life Sciences and Biotechnology

https://doi.org/10.62644/iajlb.2023.v20.i3.pp23-25

Pleural, Pancreatic and Muscular Metastasis of Dermato Fibro Sarcoma: A Report Case and Review of Literature

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ABSTRACT

Although systemic metastasis is uncommon in dermatofibrosarcoma protuberans (DFSP), this uncommon skin tumour is thought to have intermediate aggressiveness due to its frequent local recurrence [1]. A CT scan showed distant metastases to the pleura and pancreas in a 38-year-old man who had been experiencing several cutaneous nodular lesions of DFSP for six months. and psoas muscle.

Keywords Dermatofibrosarcoma, Protuberans, Metastasis, Pancreas.

Introduction

Atypical skin tumour Dermato FibroSarcoma Protuberans (DFSP) was first documented by Darier and Ferrand in 1924. DFSP is classified as an intermediate malignancy because it often shows signs of local recurrence and, less commonly, systemic metastasis [2].

Case Report

A lady who appeared with the appearance of many distinct nodules on her neck, head, and upper body was 38 years old and had no history of medical problems. Ulcerative masses was the result of the nodules' progressive enlargement.

Multiple biopsies revealed a tumour in the dermis and subcutis, with a histological analysis revealing a storiform pattern of interwoven bundles of spindle cells with plump nuclei. A diagnosis of DFSP was brought about by the biopsy.

Three additional sites were identified during the patient's pre-operative thoraco-abdomino-pelvic CT scan: a pancreatic mass with heterogeneous enhancement (Figure 2), an extensive pleural mass with compressive atelectasis of the underlying lung and invasion of the intercostal spaces (Figure 1), and a retroperitoneal mass at the expense of the psoas muscle (Figure 3).

Discussion

A low-grade sarcoma, dermtofibrosarcoma protuberans (DFSP) is a tumour of soft tissues that is intermediately malignant. Middle age (20-50 years) is the most common time for DFSPs to arise [3].

Research has linked a chromosomal translocation to the development of a fusion protein that stimulates tumour growth by increasing the synthesis of platelet-derived growth factor (PDGF), while the exact origin of dermatofibrosarcoma protuberans remains unknown. A skin biopsy is used to make a diagnosis [4].

The metastatic potential of DFSP lesions is modest, although they do recur often, and there have been isolated cases of metastases and mortality [3].

Classic DFSP is characterised by a low-grade tumour, which accounts for roughly 85-90% of cases. The risk of metastasis is relatively low, at about 5%, and the probability of local recurrence is about 26%. Approximately 58% of the remaining individuals have a fibrosarcomatous form, and about 15% will have metastasis to important organs; these patients also have a higher chance of developing local recurrence. [5].

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An incisional or excisional biopsy of the skin is the gold standard for diagnosing dermatofibrosarcoma protuberans. It is important to do a thorough history and physical examination, which should include a lymph node inspection. There is presently no widespread suggestion to use chest imaging to check for metastases before therapy, but some sites do urge it. To better characterise tumour extension before surgery, a preoperative MRI is sometimes done, albeit it is not always essential [6].

Surgical excision is the gold standard for treating dermatofibrosarcoma protuberans. Mohs micrographic surgery (MMS) is the gold standard for surgical techniques because it guarantees full histopathologic margin control during surgery. Dermatofibrosarcoma protuberans often has an unanticipated subclinical extension, making MMS a better choice than extensive local excision. It is possible to do a broad local excision with margins of 2 to 4 cm in some cases or in the absence of Mohs micrographic surgery [7][8].[9].

Oral tyrosine kinase inhibitor chemotherapeutic drugs like imatinibmesylate are an option for treating recurring,

dermatofibrosarcoma protuberans in adults, and it is not resectable. Imatinibmesylate inhibits the PDGF-beta receptor's (a tyrosine kinase) ability to bind ATP. Because of this, kinase activity is reduced, which in turn promotes apoptosis and restricts tumour development. Screening for the t(17;22) translocation should be done before starting medication since patients with this translocation respond better to imatinibmesylate. To check whether the translocation has taken place, one may use either RT-PCR or fluorescent in situ hybridization (FISH). The imatinibmesylate side effect list includes gastrointestinal distress, swelling, lethargy, anaemia, and rash. According to research, over 65% of individuals with dermatofibrosarcoma protuberans with the translocation had a positive response to imatinibmesylate treatment. There is no set timeframe for treatment. There is some evidence that suggests a 6-month course of treatment, although this may be prolonged if necessary. For tumours that cannot be surgically removed or that return, radiation treatment is an alternative option; further radiation, known as adjuvant radiation, may reduce the likelihood of local recurrence [7][8][9].

Close clinical follow-up is necessary for patients after therapy due to the frequent occurrence of local recurrence. Recurrence is most likely to occur in the first three years after treatment; hence, patients should have evaluations every three to six months during this period and once a year after that. Due to the increased risk of local recurrence and metastasis, several sources recommend baseline and serial chest CT scans for fibrosarcomatous dermatofibrosarcoma protuberans. Unless symptoms indicate metastases, regular imaging is not necessary [6].

Conclusion

The uncommon superficial tumour known as dermofolliosarcoma protuberans (DFSP) has a low risk of metastasis but a high probability of local recurrence. In order to identify both local recurrence and metastasis, long-term radiological follow-up is essential.

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