Bahrain Medical Bulletin, Vol. 34, No. 3, September 2012

Answers to Medical Quiz

Suhair Khalifa Al Saad, MB, ChB, FRCSI, CABS, CST*
Basma Al Sayed, MB, ChB**

A1. • Dermatofibrosarcoma protuberans (DFSP).
- Carcinoma of the skin.
- Malignant melanoma.
- Keloid and hypertrophic scar.
- Epidermal inclusion cyst.
- Lipoma.
- Hemangioma.
- Inguinal hernia.

A2. The best modality of treatment is a wide local surgical excision with pathologically negative margins.

The patient had excision with free margins. The histopathology was Dermatofibrosarcoma protuberans, completely excised with free margins all around except the deep margins where femoral vessels were in close contact. Postoperatively, the patient received local radiation.

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is a locally malignant lesion reported in the literature since 1890[1]. In 1924, it was described by Darier and Ferrand and was named 'Progressive and recurring dermatofibrosarcoma or skin fibrosarcoma'[1]. Hoffman, in 1925, named it dermatofibrosarcoma protuberans[1].

DFSP is a malignant skin lesion, which originates from the fibroblast cells in the dermis and invades deeper layers like fat, fascia, muscle, bone or even blood vessels.

DFSP tumors have the chromosomal translocation t(17;22) in over 90% of cases. The collagen gene fuses with the platelet-deriver growth factor gene. The fusion gene in the fibroblast is expressed in the belief that it codes for collagen. The fusion protein is processed into mature platelet-derived growth factor which is a potent growth factor. Fibroblasts start a self-stimulatory growth signal because it contains the receptor for the growth factor. This stimulates the rapid cell division and formation of the tumor[2].

DFSP has no racial predilection and almost equal sexual distribution or a slight female predominance[3].

Patients with DFSP typically present with a small papule or patch, which is likely unnoticed at the start. The tumor gradually increases in size until it becomes a nodule, or spreads wider into an atrophic and/or sclerotic plaque. Rapid growth, ulceration and hemorrhage may complicate the picture. Usually the surface of lesion is telangiectatic.

DFSP mostly involves the trunk (42-72%), followed by the proximal extremities (16-30%); rarely it involves above the neck (10-16%)[1].

No staging system developed by the American Joint Committee on Cancer for DFSP due to its very low risk of metastasis. The disease is mostly local. A staging system was published in "Short German guidelines for clinical use.

- Stage I - Primary tumor, localized disease
- Stage II - Lymph node metastasis
- Stage III - Distant metastasis[4]

Diagnosis of DFSP is mainly clinical. Pulmonary metastasis could be detected by chest radiography. Ultrasonography may be helpful for measuring the extent of DFSP locally and for regional lymph node metastasis. Computed tomography scanning is indicated in certain conditions. MRI is helpful for preoperative assessment, to define the approximate tumor border and depth. Fluorodeoxyglucose (FDG)–positron emission tomography scanning may also be helpful in diagnosing metastatic disease.

Fine needle aspiration cytology in DFSP is not a standard diagnostic test, but reverse transcriptase polymerase chain reaction (RT-PCR) and fluorescence in situ hybridization (FISH) could be used as screening tools for the presence of COL1A1-PDGFB fusion gene before initiation of oral Imatinib molecular-targeted therapy. Imatinib is sensitive in DFSP tumors which has the classic t(17;22) translocation mutation. A skin biopsy is essential for definitive diagnosis of dermatofibrosarcoma protuberans (DFSP)[5].

Conventional chemotherapy is not used in the treatment of DFSP. Radiation therapy (RT) has been used in combination with surgery. Radiation therapy is recommended for patients if the margins of resection are positive or adequate wide excision is not possible because it may result in major cosmetic or functional deficits[6].

Author contribution: All authors share equal effort contribution towards (1) substantial contributions to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published. Yes

Potential conflicts of interest: No

Competing interest: None **Sponsorship:** None

Submission date: 30 June 2012 **Acceptance date**: 7 July 2012.

Ethical approval: Approved by Surgical Department, Salmaniya Medical Complex.

REFERENCES

1. Lemm D, Mugge LO, Mentzel T, et al. Current Treatment Options in Dermatofibrosarcoma Protuberans. J Cancer Res Clin Oncol 2009; 135(5): 653-65.
2. Sirvent N, Maire G, Pedeutour FO. "Genetics of Dermatofibrosarcoma Protuberans Family of Tumors: From Ring Chromosomes to Tyrosine Kinase Inhibitor Treatment". Genes Chromosomes Cancer 2003; 37(1): 1-19.
3. Criscione VD, Weinstock MA. Descriptive Epidemiology of Dermatofibrosarcoma Protuberans in the United States, 1973 to 2002. J Am Acad Dermatol 2007; 56(6): 968-73.
4. Ugurel S, Kortmann RD, Mohr P, et al. Short German Guidelines: Dermatofibrosarcoma Protuberans. J Dtsch Dermatol Ges 2008; 6(Suppl 1): S17-8.
5. Gloster HM Jr, Harris KR, Roenigk RK. A Comparison between Mohs Micrographic Surgery and Wide Surgical Excision for the Treatment of Dermatofibrosarcoma Protuberans. J Am Acad Dermatol 1996; 35(1): 82-7.
6. Rutgers EJ, Kroon BB, Albus-Lutter CE, et al. Dermatofibrosarcoma Protuberans: Treatment and Prognosis. Eur J Surg Oncol 1992; 18(3): 241-8.