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RESEARCH ARTICLE

DERMATOFIBROSARCOMA PROTUBERANS OF THE MONS PUBIS - A RARE CASE REPORT

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is an uncommon soft tissue tumour that involves the dermis, subcutaneous fat and in rare cases, muscle and fascia.The tumour typically presents as a slowly growing, firm plaque on the trunk of young adults. It has intermediate malignant potential with a tendency for local recurrence. The treatment generally involves wide local excision with negative margins. It rarely involves the vulva and not many such cases have been reported in literature. In this case report, the authors describe a case of DFSP involving the mons pubis in an elderly female.

Introduction:-

Dermatofibrosarcoma protuberans (DFSP) is an uncommon low-grade tumour of the skin and subcutaneous tissues. DFSP was first described nearly a 100 years ago by Darrier and Ferrand in 1924, with the currently accepted term proposed by Hoffman in 1925 (1, 2). The incidence of DFSP is 0.1% of all cancers and 1% of all soft tissue sarcomas (3). It most commonly involves the trunk or proximal extremities and is generally seen in young to middle aged adults. DFSP is a rare fibrous tumour which is slow growing, superficial with intermediate malignant potential and is often locally aggressive with high rate of local recurrence (4). DFSP may develop a high-grade fibrosarcomatous component (5). It rarely involves the vulva. Primary sarcomas of the vulva would make up around 1-2% of all vulvar malignancies (6). The most common clinical presentation is a firm plaque with surrounding red to blue discoloration, or less often with multiple small subcutaneous nodules (7). Herein we report an unusual case of this tumour involving the mons pubis in an elderly patient.

Case presentation:-

A 68 year old post-menopausal female with no known comorbidities presented to our OPD with the complaints of recurrent swelling over the vulva over the last decade. The patient was initially evaluated in 8 years ago in an outside hospital where she had presented with a painless nodule over the vulva for which she was operated. We do not have the details of the surgery or post op histopathology of that time. The patient again presented in an outside hospital 4 years ago with the similar complaints. She underwent wide local excision and the post-operative histopathology showed spindle cell tumour of intermediate malignancy suggestive of DFSP. The patient did not receive adjuvant treatment at that time.

The patient presented to our OPD with the complaints of swelling in the vulva for the duration of 4 -5 months. It was insidious in onset and gradually progressive in size. It was associated with pain which was continuous and non-

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radiating. On Examination there was a solitary swelling about 4 x 4 cm in the mons pubis. It wasnon-tender, fixed with diffuse margins and overlying skin was free.

Her ultrasonography showed one large oval hypoechoic solid mass in lower part of mons pubis near upper junction of labia majora. Her chest X ray was normal.

After discussion in multidisciplinary tumour board she was taken up for Wide Local Excision. On op 5 x 4 x 2 cmmass was seen near mons pubis – anterior commissure which was mobile and was not adherent to underlying structures.

Post-operative Histopathology showed a malignant spindle tumour present in the dermis and subcutaneous tissue and overlying epidermis was atrophic. Tumour cells were spindle shaped, running in bundles and forming storiform or hering bone pattern. Tumour cells showed moderate pleomorphism and elongated hyperchromatic nuclei. Mitotic figures were frequent in large areas and surgically resected margins were free with the final impression of dermatofibrosarcomaprotuberans – high grade. Immunohistochemistry was positive for Vimentin and CD 34.

The patient then received Post-operative external beam radiotherapy to the tumour bed using electrons to a dose of 60Gy in 30 fractions over a period of 6 weeks. The patient is on follow up since 1 year with no evidence of recurrent disease.

Discussion:-

DFSP is a rare and slow growing tumour of cutaneous origin. The natural history of the disease is characterized by slow, infiltrative growth over years and decades (3). However, it can be locally aggressive and it may require multiple excisions. This was the scenario with our patient as well. The patient had history of previous 2 excisions over an eight year period prior to the resection done at our centre. Approximately 1%–4% of patients will develop distant metastasis, typically many years after the initial lesion develops (8). Distant spread of disease is believed to arise from the high-grade fibro sarcomatous component (5). DFSP is not characterized by a clear racial or gender predilection and can occur across the age spectrum, with congenital DFSP being rare (9). Grossly, the tumour may have a plaque or nodular appearance, which may be red or skin-colored, with surrounding red or blue discoloration. It may be solitary lesion but also may present with multiple foci (10). In our case, the patient presented with a solitary nodular lesion over the mons pubis.

On histopathology DFSP has more of storiform pattern with little nuclear pleomorphism. Necrosis is usually absent (11). On Immunohistochemistry DFSP is characterized by presence of CD34 (12). The expression of CD34 support the view that this malignancy is a variant of nerve sheath tumour.

DFSP is characterized by a chromosomal translocation between distinct regions of chromosomes 17 and 22, t(12;22), leading to fusion of the collagen 1 alpha 1 (COL1A1) gene to the platelet-derived growth factor B (PDGFB) gene (13). The resulting fusion protein COL1A1–PDGFB is processed to yield fully functional PDGFB. It is thought that this autocrine PDGF receptor stimulation contributes to the development and growth of DFSP (14).

Standard treatment of DFSP is surgical excision. It involves wide local excision extending 3-5 cm beyond the margins (15). Local recurrences in up to 20- 60% of cases have been reported when margins are inadequate (16). However, when margins are clearly negative, the local recurrence rate is significantly lower, with 4% at 10 years reported (17). Some physicians have advocated Mohs' micrographic surgery as the primary means of resection; however, this technique is not routinely available and has not been directly compared with wide surgical excision alone (18). In ower recurrence rates have been reported with MMS (19). MMS allows mapping of the tumour along with microscopic examination of various margins and aids in tissue preservation (20). The safety margin of DFSP resection is the hotspot because of its infiltrating growth pattern.An adequate surgical margin remains the key to reduce the recurrence of DFSP.

Radiation has occasionally been used as a primary therapeutic modality but it is commonly used as adjuvant therapy after surgery. Post-operative radiation therapy is a preferred option for positive or close surgical margins if further resection is not feasible (21). In our case, since it was a recurrent tumour, it was decided to treat the patient with radiotherapy. Recently, a meta-analysis reported that patients undergoing postoperative radiotherapy had a lower recurrence rate compared with those undergoing surgery alone (22).

In a phase II study, there is evidence of role of molecular therapy with imatinib in cases of DFSP with good response rates (23). Imatinib is an option for tumours that overexpress PDGFRB. It is an option in metastatic disease or in locally inaccessible diseases as well.

Conclusion:-

In summary, DFSP is an interesting soft-tissue neoplasm of the subcutaneous tissues with a unique chromosomal translocation that drives its growth. Wide surgical excision remains the mainstay of treatment in patients with resectable and localized disease.

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