DERMATOFIBROSARCOMA PROTUBERANS --A RARE VARI^ANTOF SOFT TISSUE SARCOMA OF SKIN. AN EXPERIENCE OF TWO CASES AT AYUB TEACHING HOSPITAL COMPLEX (ATHC), ABBOTTABAD

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(*For extreme disease, extreme method of cure)*

CASE:1

An old lady of 58 years' age presented with a huge soft tissue mass over left pectoral region which was gradually increasing in size over the last 5-ycars. Patient also gives history' of recent appearance of constant dull pain and low grade fever (<100F°) for the last 3-4 months. CT Scan revealed a soil tissue sub cutaneous mass extending from the dermis downwards without rib erosion. X-Ray chest clear and USG abdomen was normal. Excisional biopsy revealed Dermatofibrosarcoma protuberans. After wound healing, external radiation on 10Mev electron beam (Varianclinac-20) with direct anterior field encompassing nearly upper 2/3 of left chest wall and delivering a tumor dose of 48 grays at I cm depth in 16 treatments. 3 'A weeks' time as ϵ 300 cgys daily.

The tumor bed can be radiated on cobolt -60 (Mega voltage beam) with tangential fields to mid line dose of 60 grays in 30 fractions for 6 weeks' time (if facility of linac is not available).

Fig-1 X-Ray of thigh showing soft tissue mass (case II)

CASE-2

A middle age women of 40 years, presented with painless soft tissue swelling over antero-lateral aspect of upper 3rd of left thigh with gradual increase in size for the last 3 ½ year. On examination there was a firm, non-tender, hardly mobile (from side to side) soft tissue mass measuring approx. 5cm in its greater diameter and over lying skin of normal colour. Accessible lymph nodes were not palpable, fiver and spleen were not palpable. C.T scan of thigh revealed a soft tissue s/c mass with increased vascularity without involvement of underlying bone (femur). Ultrasonography abdomen was normal and x-ray chest clear. Excisional biopsy revealed Dermatofibrosarcoma protuberans. External radiation was planned on cobolt *60* with lateral opposing fields to midline dose of 60 grays in 30 fractions *for 6* weeks' time.

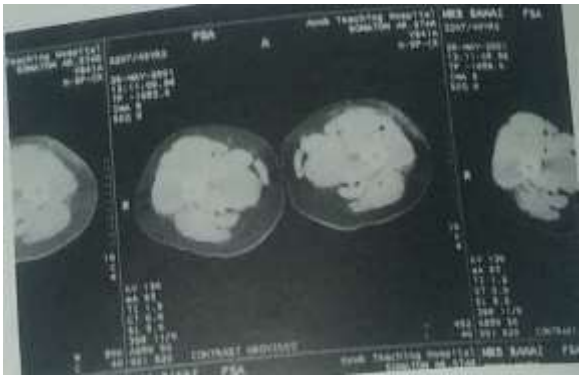

Fig – 2 CT Scan Showing Soft tissue distortion (Case – II)

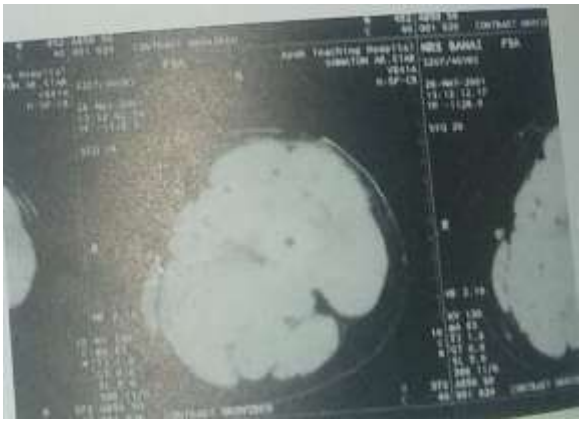

Fig – 3 CT Scan Showing Soft tissue distortion (Case – II)

DISCUSSION

Dermatofibrosarcoma Protuberans (l)ISP) is an uncommon locally malignant slowly growing tumor originating in the dermis from fibroblast and histogenecally linked to histiocytes and labeled as malignant fibroblastic histiocytoma of the skin [2,13 12]. It appears clinically as small indurated dark red colour firm nodule or fibrous plaque on one side of midline usually on trunk and proximal extremities and rarely on head and neck region[4567]. Patients are usually middle aged complaining of a firm painless lump (nodule) in the skin that has been slowly increasing in size for several years. These lesions remain intra-cutaneous until the plaques coalesce and begin to grow rapidly and if left untreated the skin surface become ulcerated with a fun gating neoplasm. Males are effected 'more than females (4:1) and it is commoner in blacks than whites[8].

Surgical treatment is associated with frequent local recurrence (over 5o%) and in many ways this tumor behaves like desmoid except that distant metastasis occasionally occur (5%)[9,10,7]. Although, some authors labelled DFSP as benign, its loco-regional behaviour is truly malignant. Treatment should be aggressive and include wide resection followed by post-operative radiation[11].

Microscopically it is a highly cellular lesion difficult to differentiate from sub-cutaneous fibrosarcoma[12] that occurs most commonly as non-specific nodules on extremities with normal skin overlying. Pigment laden cells may occur and these lesions predominantly affect persons of colour called “Bednar tumors”[13,14]

Electron microscopic studies favour a neuro-fibroblastic origin and tissue culture experiments surest histiocytic origin [5,6]. Many general surgeons lack experience in the management of DFSP and this probably accounts for the performance of sub- optimal surgical procedure in many patients especially at DHQ Hospitals. Even in teaching hospitals, the routine practice is simple excision (enucleation). This procedure involves removal of the gross tumor with the Pseudo-capsule where microscopic extension remains in-Situ and tissue planes are often contaminated by the procedure and the resulting Hameatoma. By itself this procedure is inadequate therapeutically and almost always results in local recurrence[17,18] in patients in whom even, extensive surgical procedures including Mohs surgery result in sub-optimal margin of resection [19, 14].

Adjuvant radiation therapy may improve local tumor control and *eradicate* sub-clinical (microscopic) disease. The *tendency of DFSP* to permeate deep into the subcutaneous tissue far beyond the site of origin makes the use of adjuvant radiation therapy particularly appealing.

Radiotherapy can be applied pre-operatively. intra-operative by and postoperative by[25 26] Experience with pre-operative radiation therapy is limited because the radiation therapist usually does not see these patients until after a surgical resection has been performed. Local control can be achieved in > 85% cases treated post operatively to doses in the range of 60-65 grays in 6- 7 wks time with satisfactory functional results. Local failure following postoperative radiation therapy is rare in patients with limited size lesion (stage-1 *& II)*, there may be increased risk of local recurrence when doses < 50 grays arc utilized.

Kiel & Suit have suggested doses >. 60 grays may be associated with greater chance of local control" but we have generally used in our practice doses of 55-60 grays with encouraging results. Patients with in-operable or un-resectable tumors have been treated with external beams o i/v radio sensitizers with good palliations[28]. Local control is quite respectable in patients with small lesion adequately' treated to full tumoricidal doses of radiation[29] but it remains essential to remove the tumor with a margin of normal tissue.

The arguments for pre-operative radiation therapy include:

i. That pre-operative treatment produces partial regression of the tumor resulting in less extensive surgical resection.

ii. It may decrease the risk of auto transplantation of the tumor in the surgical bed and of intravascular seeding.

Atkinson and associates’[0] reported excellent local control following moderate doses 45- 50 grays in.4-5 weeks’ time and block resection. Martin and co-workers have reported similar results in patients with advanced lesions treated with 50-70 grays pre-operatively. Suit *et a/.,* showed an actual local control rate of 89% at 5 years [32,34] McNeer *et al.* (Memorial Hospital NYC) reported 57 % 5 years’ survival with adjuvant radiation therapy[35]. Pre-operative radiotherapy plus hyperthermia may also have promise.[36]

Many surgeons and radiotherapists favour post-operative treatment. Post-operative radiotherapy must be delayed until adequate wound healing has occurred.

CONCLUSION

It has become clear that in most patient’s radical surgery resulting in mutilation can be obviated by the judicious use of adjuvant radiotherapy. The optimal combined modality approach is still under investigation.

The extent of surgical procedure, timing and adequate doses of radiation, use of Electron beam of varying penetration, sparing of medial or literal strip of tissue to provide lymphatic drainage in the extremity and proper selection of treatment aids such as beam shaping blocks, tissue equivalent bolus, missing tissue compensator and wedge filters for beam hardening and immobilization devices for patients positioning are not only essential but highly desirable and must be defined more precisely.

Several randomised trials have shown no benefit to chemotherapy in these tumors and its potential value must be weighed against the significant toxicity.

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