Dermatofibrosarcoma protuberans (DFSP) is a rare form of soft tissue sarcoma that tends to invade and recur locally. The most common site of this disease is head, neck and extremities; however, the involvement of the breast has been reported. We present a case with a dermatofibrosarcoma of the breast that is composed of low-grade spindle cells which are positive for CD45. She was treated by wide surgical excision followed by adjuvant external beam irradiation at a dose of 60 Gray. After a follow-up of 24 months, no local-regional or distant recurrence was detected.

Keywords: Dermatofibrosarcoma, DFSP, dermatofibrosarcoma protuberans, dermatofibrosarcoma of the breast, radiotherapy in dermatofibrosarcoma

The tumour bed was identified, contoured and 3 centimetres margin is added to form the Clinical target volume receiving 60 Gy; CTV-60Gy (Figure 5; the generated CTV). An additional margin of 10 mm is added to create the PTV to avoid mistreatment due to setup errors. The physics plan was simple; just two opposing large fields and two small fields in fields, physical wedges were used to improve dose distribution (Figure 6; beam arrangements and PTV coverage by 95% of the dose). According to the DVH, 95% of the PTV is covered by 95% of the dose. The liver, lungs, right and left ventricles’ mean doses are far below the recommended limits; V20 of right lung is limited below 10%, V25 of bilateral ventricle less than <5%.

After 24 months of follow up, no local regional recurrence was detected, however, she complained from dark skin discoloration and edema at the site of the radiotherapy (figure 7; edematous changes within the operative bed post radiation).
lymph node dissection failed to show any benefit in improving the recurrence rate only if it were used by experienced surgeons (10).

techniques such as Mohs surgery are acceptable to reduce the recurrence to that, adequate margin from the skin, subcutaneous tissue not less than 2-3 centimetres across all tumour dimensions. In ad-

The pathological diagnosis of DFSP depends upon histopathological examination and immunohistochemistry (IHC) of the surgical specimen. This tumour usually characterized by being formed of spindle cells arranged in a storiform pattern with little nuclear pleomorphism and mitosis. DPFS are usually CD34 positive in 90% of the cases and negative to other markers such as S-100, actin and desmin (6).

Few publications have suggested a relationship between up-regulated PDGFB gene and occurrence of DFSP. This upregulation occurs secondary to the translocation between collagen type 1 gene located on chromosome 17 and platelet-derived growth factor B-chain located on chromosome 22 (3). Although, this translocation failed to predict the behaviour and mitotic potential of the disease, one publication suggested that the use of CD34 and D2-40 might do that role (6, 7).

Since DFSP is known to be chemotheraphy and radiotherapy resistant, the main cornerstone of therapy is surgical resection with an adequate margin (8). DFSP tends to spread within the subcutaneous tissue and the underlying fascia and muscle, hence, it has high local recurrence rate, especially within the first three years post-resection (5). Despite that, it is still unknown the exact margin needed for the resection. It is widely accepted to excise the disease with margin not less than 2-3 centimetres across all tumour dimensions. In addition to that, adequate margin from the skin, subcutaneous tissue and fascia must be included in this resection (8, 9). The new novel techniques such as Mohs surgery are acceptable to reduce the recurrence rate only if it were used by experienced surgeons (10). Elective lymph node dissection failed to show any benefit in improving the local recurrence rates (2). Several factors have been linked to the increased risk of local recurrences such as positivity of surgical margin and Ki67-index (11).

The use of adjuvant external beam irradiation has been linked to an improved local control following the excision, especially in those with adverse prognostic factors such as high Ki67 or positive margin (2, 12). Still, it is not well-known the recommended dose of radiotherapy in DFSP, however, many authors reported dose range between 55.8 – 66.0 Gy in standard fractionation, 1.8 – 2 Gy per fraction, in the adjuvant setting, however in the neoadjuvant setting the range was between 50 – 50.4 Gy (13, 14). Although these authors reported a local control rate post-radiotherapy up to 98% and 95% at 5 years and 10 years, these studies lacked the minimal requisites to accept them as strong medical evidence in favour of radiotherapy (14). They lacked the presence of comparative arms as well as appropriate randomization (15). In general, the adjuvant radiotherapy is an acceptable option for those with adverse factors to improve the local control.

Although several authors reported a case of DFSP in the breasts, this case remains to be unique since it represents a successful case that is not only treated by wide resection but also by adjuvant radiotherapy (5, 16-19). Despite the narrow resection margin with less than 1.5 cm, the use of adjuvant external beam irradiation, 60 Gray, succeeded to prevent local recurrence for the entire 24 months of the disease follow up.

Dermatofibrosarcoma is a rare neoplasm that originates from the fibroblast within the subcutaneous and fascial zones. Although it is quite common to see this neoplasm in the head, neck and extremities, it is uncommon to see it in the breast. Till now, no specific guideline recommendation for dealing with DFSP of the breast, however, current management is based on surgical excision with safety margin more than 2-3 cm beside adjuvant radiotherapy in case of narrow margin, recurrence or high mitotic index. This paper presents a case of DFS of the breast that had wide excision but with safety margin less than 1.5 cm followed by adjuvant external beam irradiation at a dose of 60 Gray to reduce the risk of recurrence. After 24 months of follow-up, the patient has no distant or loco-regional recurrence.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

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References


