DERMATOFIBROSARCOMA PROTUBERANS OF THE ANTERIOR ABDOMINAL WALL: A CASE REPORT

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ABSTRACT
Dermatofibrosarcoma protuberans (DFSP) is a slowly growing neoplasm with low metastatic potential; occasionally it may attain large size. Case presentation: We report a 8cm*6cms anterior abdominal wall tumor in a 55yrs old female patient with no previous history of surgery. Preoperative CT Scan demonstrated a large tumor arising from the subcutaneous tissue of anterior abdominal wall in right hypochondrial region, lying just superficial to rectus abdominus muscle, with fat planes maintained. Subsequent histology revealed a dermatofibrosarcoma tumor. Conclusion: Surgery with negative resection margins remains the treatment of choice. This case is worth presentation because of its uncommon site of occurrence.

Key Words: Soft Tissue Tumor, Anterior Abdominal Wall, Surgical Treatment, Abdominal Wall Reconstruction

INTRODUCTION
Dermatofibrosarcoma protuberans is a rare multinodular subcutaneous or dermal neoplasm of fibrohistocytic origin commonly arising on trunk or extremity. It affects either sex but occurs slightly more often in men. It has higher incidence between the age group of 30 & 50 yrs (Daniel et al., 2004). Dermatofibrosarcoma protuberans is a low-grade sarcoma arising from dermal fibroblasts. The lesion appears as a smooth nodule in or immediately beneath the skin (trunk 40% and head/neck 40%) in mid-adult life. Due to their slow growth, lesions are commonly 1 to 2 cm at diagnosis. The external appearance belies the true character because tumor cells frequently invade the underlying soft tissues, leading to incomplete excision and local recurrence. Treatment consists of WLE with 2- to 3-cm margins (Kourtney et al., 2004). Soft tissue sarcomas are mesenchymal neoplasms comprising 1% of adult malignant tumors. Among soft tissue tumors, abdominal wall tumors are uncommon, accounting for less than 5% of these neoplasms (Brennan, 1999; Khatri et al., 2003). Most common soft tissue tumor of the abdominal wall is the desmoid tumor while the least frequent is dermatofibrosarcoma protuberans (Stojadinovic et al., 2001). Distant metastases are uncommon and are preceded by two or more local recurrences. Radiation therapy has been used effectively after resection of recurrences or for remnant disease (Cournet et al., 2004). More than 90% of DFSP feature a translocation between chromosome 17 & 22. The growth of DFSP is a result of deregulation of PDGF beta chain expression & activation of PDGF receptor protein tyrosine kinase (De Vita and Hellman, 2008). The typical presentation is that of a painless enlarging mass. Despite their benign histological appearance they show aggressive local behaviour. Treatment of these tumors is wide local excision with a tumor free margin. The local recurrence rate approaches only 10% if margins are tumour free (De Vita and Hellman, 2008). Fortunately, systemic metastases are rare. DFSP is a Radiosensitive tumour. So it is an option where we have to do conservative excision, where it is followed by radiotherapy.

CASES
A 55-year-old female presented with a slowly growing mass in the rt hypochondrium, gradually increasing in size with multiple dark pigmented nodules (size ranging from 2mm to 10mm) appearing on its surface. There was no history of trauma or operation for any cause in the past. It started as a painless
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mass 2 yrs back and progressed slowly to present size. Patient noticed skin changes 6 months back. On examination, the patient had a firm mass extending in the right hypochondrium about 3 cms from midline. The mass was 8*6 cms and was mobile, non tender. It became prominent on straight leg raising test. The computed tomography scans showed a solid skin-subcutaneous lesion not invading the underlying fascia or musculature. Intraoperatively, the tumor was invading the superficial layers of skin as seen from inside. The underlying muscle was not infiltrated. Skin including all the superficial lesions and the tumor was excised with 2cms margin on all sides. The reconstruction was performed by mobilizing adjacent skin and subcutaneous tissue and primary wound closure was possible. Propylene mesh was used to close the aponeurotic defect. A closed suction drain was used. Subsequent histopathology revealed features of dermatofibrosarcoma protuberans & was negative for S 100.

DISCUSSION

Sarcomas of the abdominal wall are difficult-to-treat neoplasms. Due to their variable histological type and grade, there are many different surgical approaches. The general recommendation is to perform a wide excision with free margins. Narrow margins are related to a dismal prognosis. A wide resection with a 2-3 cm margin in the treatment of abdominal wall sarcomas is associated with a good regional and distant control. DFSP usually presents as a nodular, violet-red skin mass on the trunk and proximal extremities. It tends to present a slow growing, nodular, polypoid neoplasms and, in many cases, its symptoms are long lasting (Brennan, 1999; Courtney et al., 2004; Stojadinovic et al., 2001). Most lesions are smaller than 5 cm. The tumour can achieve an enormous size with multiple satellite nodules if left untreated (Arther and Thomas, 2011). In a large series of 159 patients treated at the Memorial Sloan-Kettering Cancer Center (New York, NY), between 1950 and 1998. Bowne et al. reported only 4 patients
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(3%) with large tumors (> 10 cm) (Lemm et al., 2009) in the present study, the authors report a DFSP that arose in an unusual site. DFSP spreads locally, with regional infiltration into surrounding structures like fascia, aponeurosis, muscles, peritoneum and bone (Khatri et al., 2003; Courtney et al., 2004; Stojadinovic et al., 2001; De Vita and Hellman, 2008; Arther and Thomas, 2011; Lemm et al., 2009; Arnaud et al., 1997). Computed tomography (CT) or magnetic resonance imaging (MRI) has been indicated to stage these tumors. These imaging techniques are important for resection planning (Brennan, 1999; Khatri et al., 2003; De Vita et al., 2004; Stojadinovic et al., 2001; De Vita and Hellman, 2008 Lemm et al., 2009). Histology shows appearance of radial whorls of spindle cells producing storiform or cartwheel pattern. Myxoid features can be present focally. Immunohistochemically they are positive for vimentin, actin & CD34. They are characterized by progressive, locally infiltrative behavior. If left untreated, these tumors continue to grow slowly, invading the surrounding tissue, including neurovascular bundles. They usually present on the trunk and extremities, between the second and fifth decades of life. Lung and regional lymph node metastases are rare and usually preceded by multiple local recurrences if limited resection is done. There is one pigmented variant of DFSP having overall appearance of ordinary DFSP with added population of dendritic cells having melanin pigment but are negative for S-100 protein. Essentially, the optimal treatment for DSFP has been a wide resection. Since these tumors are locally infiltrative, the general surgical principles used for sarcomas should be used to properly treat them. A three-dimensional wide resection of skin and surrounding structures must be performed. Most authors recommended a 2-3 cm local margin including the underlying deep fascia and overlying skin (Brennan, 1999; Khatri et al., 2003; De Vita et al., 2004; Stojadinovic et al., 2001; De Vita and Hellman, 2008 Lemm et al., 2009). When resections are performed with inadequate margins, the reported local recurrence rate can be as high as 60% (Stojadinovic et al., 2001). Recently Mohs Microscopic surgery that permits microscopic control of excised margins is advocated; it allows the subclinical margins to be mapped at time of surgery & consequently the margins more precisely determined (Arther and Thomas, 2011; Paradisi et al., 2008; Popov et al., 2007). In conclusion, when adequate principles are followed, DFSP has a good prognosis. However, close surveillance is required since local recurrence rates are high (Brennan, 1999; Stojadinovic et al., 2001). Present study showed that safe diagnosis can lead to excellent surgical results in the treatment of DFSP. The abdominal wall defect often needs reconstruction using flaps or prosthetic material.

REFERENCES


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