Dermatofibrosarcoma protuberans: Rare cause of head and neck swelling

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Abstract

Dermatofibrosarcoma protuberance of head and neck is a rare neoplasm and accounts for less than one percent of Head and Neck Neoplasm. The objective of this report is to understand etiology, presentation, histopathologic identification and treatment option for this tumour. The goal of current report is to acquaint the clinician with presentation and treatment option for this disease.

Introduction

Sarcomas in general occur rarely in Head & Neck region accounting for less than one percent of Head & Neck malignancies. Dermatofibrosarcoma of Head and Neck is a rare neoplasm. It is fibrohistiocytic tumor of intermediate malignancy that manifests exclusively on the skin. It grows slowly, is locally invasive and rarely forms metastasis. The tumor is predominantly located on trunk as well as proximal side of the extremities. It may rarely occur in Head and Neck in middle aged person. Dermatofibrosarcoma shows a tendency towards local recurrences. It occurs rarely under one per lac residents a year and average age of patient is around 40 years. Men and women are equally affected and mortality is low. We report this rare entity in a young female patient who was treated surgically.

Case report

A thirty year old Hindu female patient presented with swelling over left side of neck in supraclavicular region of eight months duration. Swelling was small to start with but over period of eight months it gradually increased to attain size of approximately 16 x 12 cm and appeared like bunch of grapes. Swelling was painless, did not bleed on touch and had no history of trauma. There was no significant history in family nor did patient have any history of tobacco chewing or smoking.

Clinically mass was approximately 16 x 12 cm in size over left supraclavicular fossa with multiple soft protuberances of varying sizes but none exceeding 5 cm, soft in consistency with thin skin over certain areas with ulceration (Fig. 1). Ulcers were infested with maggots.

There was no other positive finding on complete Head and neck examination nor there any other swelling elsewhere in the body. Systemic examination was unremarkable. Routine hematological examination and urine examination were normal except mild anemia. Renal parameters were within normal limits. Punch biopsy from ulcerated area was sent for histopathological examination which revealed diagnosis of dermatofibrosarcoma.

Cat scan of patient from base of skull to carina was done with plain and contrast studies which revealed heterogeneously intensely enhancing lesion involving skin and subcutaneous tissue in left neck and upper chest with few non specific lymph nodes in left axilla and level four lymph nodes along left internal jugular vein., findings favoring a vascularized lesion probably (a) haemangiosarcoma, or (b) haemangioma (Fig. 2).

After correcting Hemoglobin levels and taking care of maggots' patient was taken up for surgery. We did classical neck dissection from above downwards to isolate and clamp Internal Jugular vein, the lower end of which was engulfed in tumor. Whole mass with overlying skin was excised with adequate safety margin which were found to be negative on frozen section (Fig. 3).

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Fig. 1 Patient showing swelling in left Supraclavicular fossa with superficial ulceration of skin

Fig. 2 CAT scan of the patient showing tumor infiltrating soft tissues with out invasion of thoracic cavity.

Fig. 3 Tenth post operative day after excision of the tumor .Pectoralis Major Myocutaneous flap in situ

Fig. 4 Diffuse cellular tumor occupying dermis and typical pattern of subcutaneous fat (H & E) stain 10 x.

The tumor was superficial to clavicle and investing layer of deep cervical fascia. Defect of skin was repaired with Pectoralis Major Myocutaneous flap.

The recovery was uneventful and patient was discharged on tenth day. Histopathological examination of specimen confirmed the diagnosis of dermatofibrosarcoma infiltrating the underlying subcutaneous tissue. The tumor was highly cellular with monomorphic appearance and brisk mitotic activity. Giant cells were inconspicuous. There were areas of collagenization. Lymphovascular embolization and perineural invasion was not seen. The base of excision was free from tumor. Eleven lymph nodes were dissected, all of which showed reactive changes. (Fig 4)

Discussion

Locally aggressive but rarely metastasize dermatofibrosarcoma of Head and Neck is rare neoplasm. Microscopically tumor is composed of uniform, benign appearing spindle cells arranged in a star form pattern infiltrating into the dermis and subcutaneous tissue. A pigment variant the so called Bednar tumor is also known. Dermatofibroma on other hand is a non infiltrating benign dermal tumor usually less than three cms in size and can be cured by local excision while dermatofibrosarcoma is low to intermediate grade sarcoma that involves in fourteen percent cases accounting for 1.4% of Head and neck sarcoma. In Head and Neck region scalp and supraclavicular fossa are the most often involved. These tumors typically display locally aggressive course with local recurrence rates as high as 60% percent. This reflects the propensity of tumor cells to invade the local tissue with tentacle like projections within clinically normal appearing skin.

Recurrence rates appear to be higher for Head and Neck lesion up to 75% than those at other sites probably due to

limitation in surgical resection. Regional and distant metastasis is uncommon accounting for 1–4% cases.

Treatment is wide surgical excision with resection margins of negative more than three cms. The rate of recurrence is directly associated with extent of resection. Adjuvant radiation has been suggested to be of benefit in selected cases.

Conclusion

Dermatofibrosarcoma of Head & Neck is a rare neoplasm with a propensity for local recurrence when surgical margins are inadequate. When wide surgical margins of at least 2.5 to 3.0 cm are achieved, including a margin of underlying fascia, prognosis is favorable. Surgical resection is the standard of care with wide local excision. Role of Radiotherapy and Chemotherapy is undefined

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