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Myxofibrosarcoma of Head and Neck Region in a Patient with Follicular Carcinoma of Thyroid

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Abstract **Case Report**

Myxofibrosarcoma (MFS) is a malignant fibroblastic tumour with variable myxoid stroma, cellular pleomorphism and curvilinear vascular pattern. It is commonly seen in late adult life, peaks in the seventh decade and usually occurs in the lower extremities. MFS of head and of the neck is extremely rare and accounts for approximately 1% of all head and neck neoplasms. A fifty two year old lady with follicular carcinoma thyroid treated six years ago, presented with a rapidly increasing neck swelling in the left supraclavicular fossa. Fine needle aspiration cytology of the cervical node was suggestive of metastatic carcinoma and she underwent left posterolateral neck dissection. Histopathology and immunohistochemistry was suggestive of high grade myxofibrosarcoma with negative margins. She was treated with adjuvant radiotherapy and has been under regular follow up for 6 months.

Keywords: Myxofibrosarcoma, High grade, Follicular carcinoma thyroid.

INTRODUCTION

Soft tissue sarcomas of head and neck are rare mesenchymal malignant neoplasms, accounting for less than 10% of all soft tissue sarcomas. Myxofibrosarcoma (MFS) is a malignant fibroblastic tumour with variable myxoid stroma, cellular pleomorphism and curvilinear vascular pattern [1]. It is the most common soft tissue sarcoma that occurs in late adult life. Only a few cases of head and neck MFS have been reported so far [1]. We report the case of a 52 year old lady with history of follicular carcinoma of thyroid treated 6 years ago, with a neck swelling diagnosed as high grade MFS. She was treated with wide excision of the tumour and adjuvant radiotherapy.

CASE DISCUSSION

A fifty two year old lady, known case of follicular carcinoma of thyroid treated with total thyroidectomy and radioactive iodine ablation 6 years ago, presented with a rapidly increasing swelling in the left supraclavicular fossa of 2 months duration. She was on thyroxine supplements post thyroidectomy. There was no family history of any malignancy. Local examination revealed a 4x 3 cm firm, non-tender, mobile, soft tissue swelling in close proximity to the distal aspect of left sternocleidomastoid muscle. Systemic examination was within normal limits. Contrast enhanced computed tomography (CECT) scan of neck and thorax revealed a partly exophytic soft tissue lesion of size 4.6 x 3 x 3 cm involving the distal third of left sternocleidomastoid muscle suggestive of a probable nodal mass infiltrating the muscle (Figure 1 and 2). Another enhancing poorly defined soft tissue swelling was seen in the left level VI region suggestive of node/deposit. There was no evidence of any pulmonary metastasis.

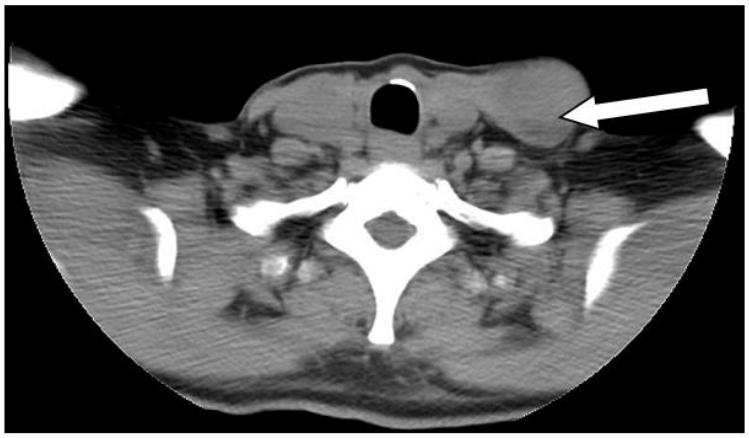

Figure 1: CT scan of neck showing exophytic soft tissue lesion of size 4.6 x 3 x 3 cm noted involving the distal third left sternocleidomastoid muscle

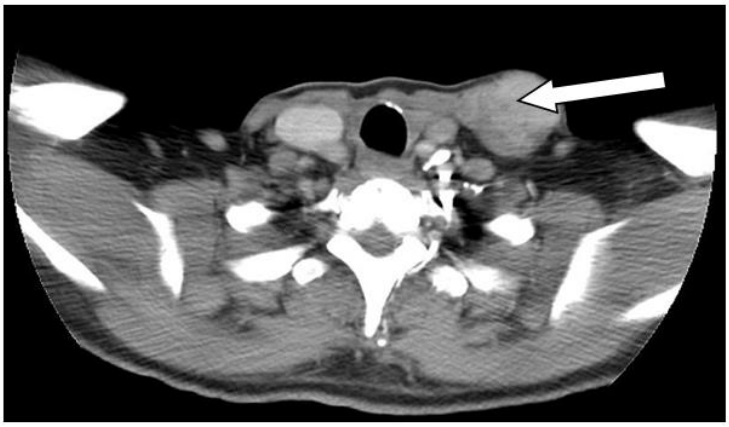

Figure 2: CECT image of neck showing enhancement of the mass lesion

She underwent fine needle aspiration cytology of the cervical node which was suggestive of metastatic carcinoma. She was investigated for metastatic follicular carcinoma of thyroid. Her serum thyroglobulin and anti- thyroglobulin antibody levels were within normal limits. In view of localised disease, she underwent left posterolateral neck dissection. Histopathological examination showed variably cellular multinodular tumour showing pleomorphic spindle cells arranged in a myxoid background with mitosis of 18 - 20/hpf (Figure 3 and 4). The tumour was seen infiltrating the skeletal muscle fibres in the periphery. Immunohistochemistry was suggestive of non-specific nuclear staining for SMA and was negative for S 100, CK and Thyroglobulin. The resected margins were free of neoplasm and seven lymph nodes dissected were reactive. The picture was suggestive of high grade myxofibrosarcoma involving part of sternocleidomastoid muscle. She received adjuvant radiotherapy to a dose of 66 gray in 33 fractions which she tolerated well. She has been under follow up since six months and has been asymptomatic.

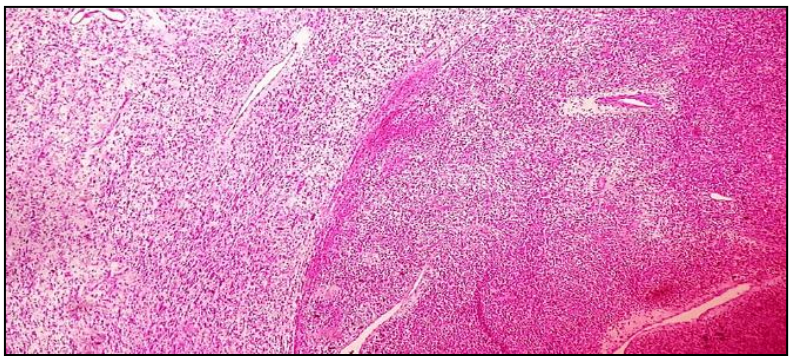

Figure 3: Variably cellular multinodular tumour showing pleomorphic spindle cells arranged in a myxoid background and curvilinear vessels seen with condensation of spindle cells around vessels

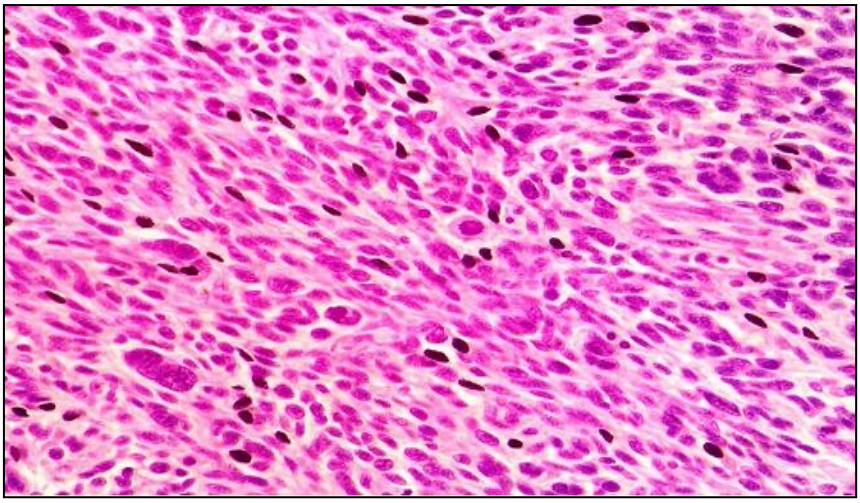

Figure 4: Hypercellular areas showing highly pleomorphic cells with increased mitosis.

DISCUSSION

MFS, the most common soft tissue sarcoma that occurs in late adult life peaks during the seventh decade. It is seen in lower extremities (77%), trunk (12%), retroperitoneum or mediastinum (8%) [1]. Involvement of the head and of the neck is extremely rare and accounts for approximately 1% of all head and neck neoplasms. MFS, previously known as a myxoid variant of malignant fibrous histiocytoma, was reclassified by the World Health Organization in 2002 and is now recognized as a distinct pathologic entity. Most common clinical presentation is a painless palpable subcutaneous lump. Skin changes on the scalp or face, subsite specific symptoms such as hoarseness, dysphagia, epistaxis, nasal obstruction or cranial nerve deficits are also seen. Rarely, MFS presents as superior vena caval obstruction [2].

Diagnosis of MFS is based on characteristic histopathologic features such as presence of alternating alternating hypocellular, myxoid areas and hypercellular fibrous areas; pleomorphic nuclei; curvilinear, thin-walled blood vessels prominent in myxoid areas; aggregation of neoplastic cells or inflammatory cells and spindle and stellate cells in myxoid matrix [3]. These tumors have been graded based on the degree of cellularity, nuclear pleomorphism, and mitotic activity [4]. Immunologic staining is typically positive for vimentin and CD-34 which indicates the tumour's fibroblastic origin, sometimes positive for SMA and negative for S-100 protein [5]. Histopathological examination in this case showed morphological features of MFS and high mitotic index (18-20 /hpf) which was suggestive of a high grade tumour. Immunohistochemistry showed non-specific staining for SMA and was negative for S-100 and CK.

Low-grade tumours rarely metastasize. They have good short term prognosis, with 5- and 10-year survival rates of 65% and 52%, respectively [6]. However, local recurrence rate is high despite repeated surgical resection with wide negative margins. Mentzel *et al*., reviewed 75 patients with emphasis on low grade tumours and documented local recurrence in 55% and distant metastases in 21% of the cases [4]. Recurrence of low grade MFS generally occurs with progression to higher histologic grades in 15–38% of cases which acquires increasing metastatic potential [7]. Sanfilippo *et al*., reviewed 158 patients with localised MFS and reported that the quality of surgical margins predicted local recurrences, whereas histologic grade determined the risk of distant metastases [8].

Surgery followed by adjuvant radiotherapy is the most common treatment strategy. A proper diagnosis followed by complete surgical resection with adequate resection margins (1–3 cm) is critical for disease cure [9]. It is often challenging to achieve adequate surgical margins within the delicate head and neck region due to anatomic constraints unlike that in trunk or extremities. Because of the low incidence of lymph node metastasis, elective lymph node dissection is not indicated unless clinically involved [10]. Adjuvant postoperative radiotherapy is recommended in patients with close (<1 cm) surgical margins and large or high-grade MFS to avoid local recurrence. Data regarding treatment with chemoradiotherapy is scarce in literature, in view of small number of cases. Cante *et al*., reported a case of MFS of maxillary sinus with complete remission at 18-month follow-up after combined chemoradiotherapy without surgery [11]. Role of systemic therapy in non-extremity sarcoma is limited. Zagars *et al*., found that adjuvant chemotherapy did not decrease metastasis or local recurrence rates [12].

CONCLUSION

Myxofibrosarcoma of head and neck is a rare tumour. Complete surgical resection with adequate resection margins is the mainstay of treatment. Nodal dissection is indicated only in case of clinical

involvement. Postoperative radiotherapy is recommended in patients with close surgical margins and large or high-grade tumours to prevent local recurrences. Adjuvant chemotherapy has no definite role.

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