

Neck exploration for a huge synovial sarcoma, case report and literature review

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Abstract

A 47-year-old woman presented in the outpatient department of EAST Surgical ward of MAYO Hospital Lahore, Pakistan, on February 2019 with complaint of swelling in the front section of the neck since five months which increased gradually in size and had been causing pain since two months. She had no comorbidities and insignificant family history. Examination revealed a 23×20 cm mass on the right side of the posterior triangle of the neck. Fine Needle Aspiration Cytology (FNAC) was inconclusive and CT of the neck showed a huge mass on the right side of the neck with cervical lymph nodes. Exploration was planned, and modified radical neck dissection Type III (Also known as Functional Neck dissection) was performed. The biopsy revealed synovial sarcoma of the neck. The patient's post-operative condition was satisfactory and she was discharged on the fifth post-operative day.

Keywords: Synovial sarcoma, Sarcoma in neck, Huge sarcoma, Functional neck dissection.

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Introduction

Synovial sarcoma is a very rare type of tumour which occurs in about 0.0001 to 0.0002 percent of individuals per year. It affects mostly men of around 30-40 years of age and male to female ratio is 1.2-1.¹

Synovial sarcoma is also known as malignant synovium and has its primary origin mainly on extremities of the body, e.g. arms, legs and sometimes close to the joint capsules and sheaths. The histology consists mainly of mesenchyme and not from the mature synovial tissue. They also have epithelial and supporting tissue features.²

However, primary synovial sarcoma is an unusual tumour of the head and neck; less than 75 cases have been reported in the literature. The prevalence of synovial sarcoma in the head and neck is about 3-10% and that is mainly from the parapharyngeal sites.³ Risk factors are still unknown but a translocation of chromosome t(X; 18)

(p11; q11) seems probable. Less than 10% of the patients had developed metastasis at the time of diagnosis. It is believed that in these patients the tumour cells moved from their original site to the metastatic site. Metastasis is more common in the lungs.³

Here we are presenting the case of a huge synovial sarcoma of the neck; surgery was performed for the excision of a huge mass on the right side of the neck with cervical lymph nodes and later it was found to be synovial sarcoma of neck.

Case Report

A female patient of 47 years of age, hepatitis C positive, presented in outpatient at the department of EAST Surgical ward of MAYO Hospital, Lahore, Pakistan, with chief complaint of painless swelling on the right side of the neck since five months. Since the last two months the swelling had increased rapidly causing pain. On examination, the swelling was 20×23 cm in size and was hard and tender (Figure-1). It deviated the trachea towards the left and encased the right carotid artery. The patient had dyspnoea while lying down. Her TSH was 6.24 mIU/L (Normal 0.25 - 4.00 mIU/L), free T4 was 16.7 mIU/L (Normal 11.5 - 23.0 mIU/L) and thyroid scan were normal. Cervical lymph nodes could not be palpated due to huge size of the mass. FNAC was done twice but was inconclusive. CT of the neck and chest showed a huge mass on the right side of the neck with solid and cystic components and positive cervical



Figure-1: Preoperative pictures of the patient showing huge neck mass.

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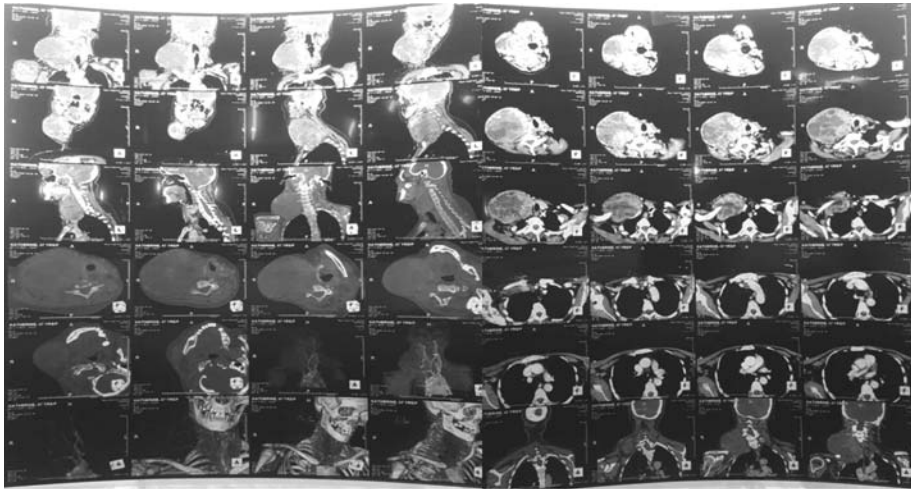


Figure-2: CT Scan of head, neck and chest of the patient.

showed glands/tubule formation while mesenchymal component showed moderate atypia. Immunohistochemical staining showed positive Cytokeratin and TLE1 in tumour cells. On receiving the biopsy report two weeks later, the patient was then shifted to the Oncology department where she was given four cycles of doxorubicin/ifosfamide chemotherapy. After eight months repeat CT scan of the head and neck was done which was clear of any residual disease.

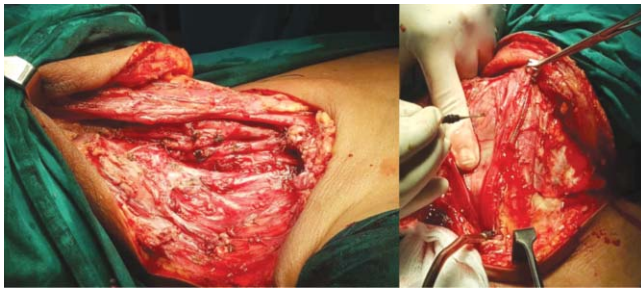


Figure-3: Intraoperative pics of the patient showing lifting of sternocleidomastoid muscle to expose the mass.

lymph nodes (Figure-2). Radiologists suggested peripheral nerve sheath tumour. Neck exploration was planned after written and informed consent from the patient along with consent for tracheostomy. Fitness for general anaesthesia was taken. Skin crease incision was made. The mass was pushing the sternocleidomastoid muscle forward. SCM was retracted and branches of external carotid artery were identified and ligated and the mass was isolated from the carotid artery (Figure-3). Modified Radical Neck Dissection Type III (Functional neck dissection) was done and the carotid artery, sternocleidomastoid and jugular vein were separated. There was no communication of the mass with trachea, oesophagus and thyroid, so thyroidectomy and tracheostomy were not performed. Suction drain was placed and surgery went uneventful (Figure-4). The patient was shifted to the ward, as she remained stable and was discharged on fifth postoperative day. The biopsy revealed Synovial Sarcoma, biphasic, FNCLCC and grade III in the posterior triangle of the neck. The microscopy showed tumour comprising epithelial and mesenchymal components. Epithelial component



Figure-4: Post-operative incision and resected neck mass.

Discussion

The name 'synovial sarcoma' was first used in the early 20th century because of its close microscopic similarity with the synovium and also due to its tendency to arise adjacent to the joints. However, the original cells of these tumours are unknown and it seems they are not necessarily of synovial origin.

Very limited data is available in literature regarding synovial sarcoma. A study by Joel et al shows data of 24 cases of synovial sarcoma, out of which 9 survived, while the rest died by pulmonary metastasis.⁴ Another study by Rossario et al shows that the biological activity of synovial sarcoma which occurs in the head and neck and those which occur on other sites are the same.⁵

The primary origin of synovial sarcoma are mostly soft tissues near the large joints of upper limbs and legs but they have been documented in various other sites of the body, e.g. brain, prostate and heart. Two types of cells are found microscopically: fibrous type and epithelial type. Fibrous type or spindle or sarcomatous cell are small and uniform and are found in sheets. Classical synovial sarcoma is biphasic with both cells present. They can also appear to be poorly differentiated, consisting of sheets of spindle cells. Few pathologists have reported that, though rare, there can be monophasic epithelial form which makes it difficult to diagnose these cells.

Just like other soft tissue sarcoma there exists no grading system to report the histopathological features of these synovial sarcomas. NCI grading system is more common in the United States, while in Europe the Trojani or French system is popular.⁶ In the Trojani Scoring system, tumour differentiation, mitotic index, and tumour necrosis are seen and score is given between 0 to 6 and which are converted into grades from 1 to 3 with 1 being less aggressive tumour. NCI system also consists of three grades but it consists of other factors as well.

The treatment of synovial sarcoma consists of surgery, chemotherapy and radiotherapy. Surgery is the mainstay of treatment and is aimed to remove the tumour and is curative in 20-70% of the patients. After the surgery, chemotherapy is used to reduce the load of micro metastasis. However, the role of chemotherapy in synovial sarcoma is unclear. Some studies have shown that in poorly differentiated disease the survival of patients is

improved with Doxorubicin/Ifosfamide treatment.⁶

To reduce the role of local recurrence, radiotherapy has its role; however, the benefit of radiotherapy is less clear than chemotherapy. Prognosis of synovial sarcoma depends on the quality of surgery received by the patients and the tumour characteristics i.e. tumour size, local invasiveness, histological subtypes, presence of metastasis and lymph node involvement. Small tumours can be completely removed with adequate margins and this gives good prognosis. Tumours greater than 5 cm are at increased risk of developing distant metastasis.

Conclusion

Synovial carcinoma in the neck is a rare tumour found in the head and neck region. Because of its rarity no clear relation is found between its occurrence at unusual site e.g. neck. In this case report, the biopsy turned out to be synovial sarcoma of the neck in a 47 year old woman which was excised successfully and patient was symptom free completely after chemo therapy. On the one hand, this case report will help in identifying this rare type of disease and that it is completely curable by surgery and radiotherapy while on the other hand it will help us in finding the cause of the tumour at these unusual sites.

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Conflict of Interest: None to declare.

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