
Synovial Sarcoma of Pyriform Fossa: A Rare Case Report**Syed Umar Shah¹, Shubha Verma², Asif A. Wani³, Masood H. Kirmani⁴, A. R. Khan⁵**¹Department of Otorhinolaryngology, SKIMS Medical College, Srinagar²Department of Otorhinolaryngology, SKIMS Medical College, Srinagar.³Assistant Professor, Department of Otorhinolaryngology, SKIMS Medical College, Srinagar.⁴Professor & HOD, Department of Otorhinolaryngology, SKIMS Medical College, Srinagar.⁵Ex- HOD, Department of Pathology, Government Medical College, Srinagar.***Corresponding author**

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Abstract: This is a case report of synovial sarcoma of left pyriform fossa in a 35 year old male who presents in ENT Department with complaints of foreign body sensation since two months and difficulty in swallowing solid foods since 1 month duration. A globular swelling was seen in left pyriform fossa on indirect laryngoscopy. A CECT neck confirmed a well defined localised mass of 2cm x 1.4cm. Excisional biopsy was taken on direct laryngoscopy. Histological diagnosis of biphasic synovial sarcoma was confirmed by immunohistochemistry and molecular analysis. Clinical follow up with CT 3 months after operation has not shown any residual disease and patient is on regular follow up. Synovial sarcoma is a rare malignant tumor. It derives from a mesenchymal precursor stem cell that is unrelated to mature synovial tissue. Synovial sarcoma classically affects lower limbs between the ages of 15 and 40 years and the proportion of male-to-female patients is 3:2. It is very rare in the head and neck region especially in laryngopharynx. We report a case of pyriform fossa synovial sarcoma confirmed by histopathology and immunohistochemistry. To the best of our knowledge this is the third case in India and first in Valley of Kashmir (Srinagar) documented in English Literature.**Keywords:** synovial cell sarcoma, malignancy, immunohistochemistry.

INTRODUCTION

Synovial cell sarcoma, or synoviosarcoma, (SS) is a mesenchymal malignancy that is termed SS since its histological appearance is similar to that of the synovium. However, SS rarely exhibits a synovial structure and is considered to originate from pluripotent mesenchymal cells. The characteristic biphasic pattern of SS is due to the two morphologically distinct but histogenetically related cell types that compose the sarcoma. Depending on the relative prominence of the two cell populations and the degree of differentiation, these tumors form a continuous histopathological spectrum of biphasic, monophasic fibrous, monophasic epithelial and poorly differentiated (round-cell) types[1,2].

Diagnosis is made on FNAC, histopathology supported by immunohistochemistry and molecular analysis.

Synovial sarcoma is a rare, malignant neoplasm that accounts for approximately 8% of all soft tissue sarcomas. It can occur at any site and age but primarily arises in the para-articular areas of the lower extremities of young adults, especially around the knee and ankle. Since the first case of head and neck synovial sarcoma reported by Jernstrom in 1954, only

3% to 5% of all cases were found in the head and neck region. In this region, the hypopharynx is the most common site[3].

We document a rare case of synovial sarcoma of pyriform fossa mass presented in our ENT & Head & Neck Surgery Department and review the literature related to this rare entity.

CASE REPORT

We submit a case of a nonhypertensive on diabetic male aged about 35 years who presented to our ENT opd with complaint of foreign body sensation throat since 2 months and difficulty in swallowing solid food since 1month. He also gave history of occasional smoking. A thorough ENT and head and neck examination was conducted, and on Indirect Laryngoscopy a globular swelling was seen in left pyriformfossa. A CECT neck was done which showed a well defined left pyriform fossa mass of about 2cms by 1.4 cms with no local extension (shown in the photograph below 1-4). Small level II and Level III nodes seen on bilateral sides. The findings of indirect laryngoscopy were confirmed by Direct laryngoscopy and excision biopsy with surrounding normal tissue was done and was sent for histopathology and immunohistochemistry.

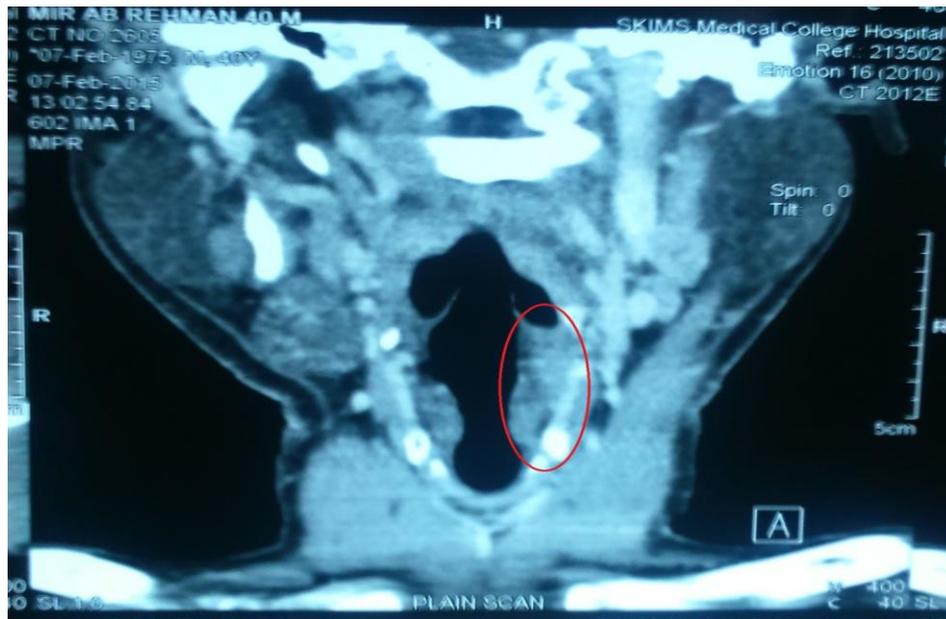


Photo-1

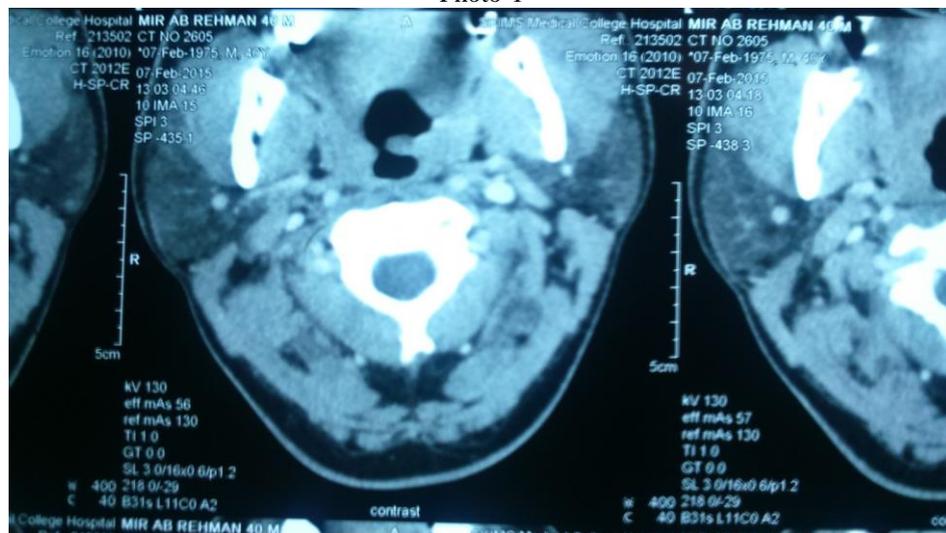


Photo-2



Photo-3

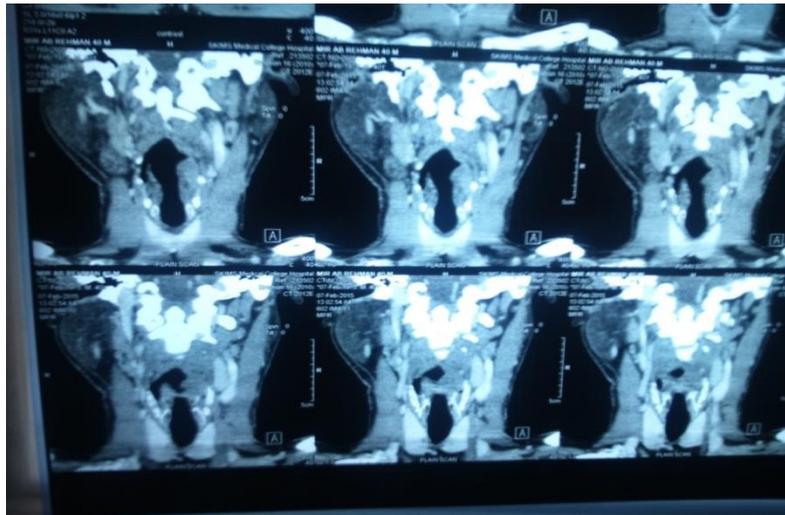


Photo-4

Photos (1-4) :A CECT neck showing a well defined left pyriform fossa mass with no local extension

Histological examination of the tissue showed a biphasic synovial sarcoma (see figure 1) composed of sheets of uniform spindle cells with deeply stained nuclei and scanty ill defined cytoplasm , epithelial component arranged in tubules lined by cuboidal cells

with uniform vesicular round nuclei and well defined cell cytoplasm as well as pale clusters of cells interspersed with spindle cells. Raw mitosis was seen. Necrosis was absent.

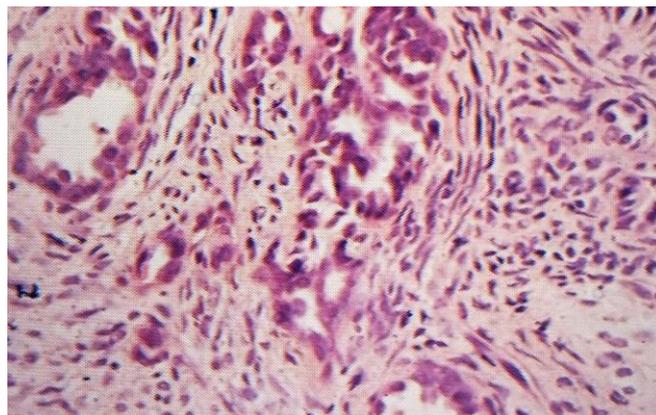


Fig-1: Histological examination of the tissue showed a biphasic synovial sarcoma

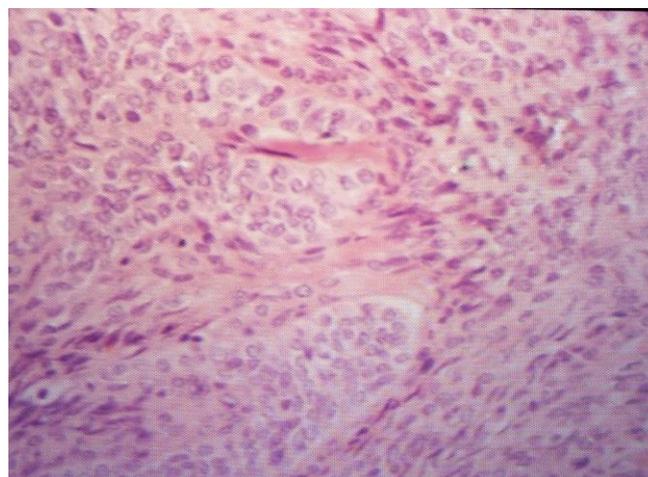


Fig-2: Immunohistochemistry showing ,glandular epithelium was positive with pancytokeratin AE1-AE3. Spindle cells express Bcl-2. Mic-2 and Calponin were expressed in both stromal and glandular elements. S-100 and CD10 were negative.

On immunohistochemistry, glandular epithelium was positive with pancytokeratin AE1-AE3. Spindle cells express Bcl-2. Mic-2 and Calponin were expressed in both stromal and glandular elements. S-100 and CD10 were negative.(fig.2)

Molecular analysis revealed a translocation SYT gene on chromosome 18 and SSX1 gene on X – chromosome.

A post biopsy MRI scan was done which showed no residual disease and patient is on regular follow ups since 1 year(as shown in picture below).(photo 5)

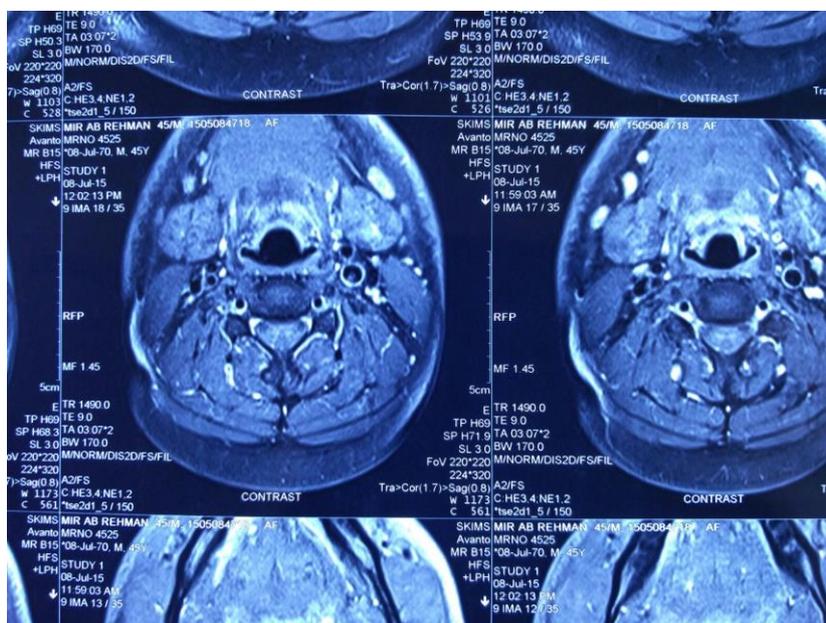


Photo 5: A post biopsy MRI scan was done which showed no residual disease and patient is on regular follow ups since 1 year

DISCUSSION

Despite its name, synovial sarcoma rarely originates in the synovial membranes. It is most commonly found in the vicinity of large joints. Occurrence in the head and neck, a location poor in synovial tissue, is uncommon. Synovial sarcoma comprises 8 to 10% of all soft tissue sarcoma and it is estimated that 3 to 10% of sarcomas occur in the head and neck .Synovial sarcomas show a male predominance (3:2)[4]. This tumor is most prevalent in adolescents and young adults aged 15–40 years, but it has been described at practically all ages[5].

SS is generally considered a malignant tumour, with an aggressive behaviour and with high probability of recurrence; in the head-neck region. The neoplasm manifests as a indolent growing mass, the symptoms of which are correlated with the site of onset which, in decreasing order of frequency, comprises hypopharynx, neck, face, larynx, rhino- and retro-pharynx. It usually occurs as a well-circumscribed mass covered by a thin fibrotic capsule making it easy to enucleate from the surrounding tissues. This is, however, a pseudocapsule beyond which the neoplasm usually tends to infiltrate. Invasion of the tumoural pseudocapsule is usually accompanied by onset of distant metastases also in very distant districts. For this reason, simple enucleation is

followed by recurrence in 90% of cases and, in radical exeresis of sarcomas of the limbs, free borders of at least 5 cm are necessary which, for anatomical limitations, are not feasible in the head and neck district .This probably accounts for the high rate of local recurrences after limited exeresis. CAT and nuclear magnetic resonance (NMR), whilst not specific, are useful in planning the surgical strategy. In 30-60% of SS cases, calcifications have been found at CAT examination and these, when present, are to be considered a positive prognostic factor. The factors associated with a worse prognosis include: age>25 years, tumour size >5 cm and low grade of differentiation[6]. However a recent study has stated that tumour size greater than 5.0 cm in diameter was an independent adverse prognostic factor for overall survival[2].

Metastasis to regional lymph nodes of synovial sarcomas in the head and neck is not seen often, although there can be clinically enlarged lymph nodes. Most metastasis originate from hematogenous dissemination, although up to 20% spread through the lymphatics to regional lymph nodes[4]. Most frequent site of metastases is the lung (49%), followed by skeleton (24%), liver (14%) and brain (11%)[6]. Cervical lymph node dissection is not routinely

performed, except in the presence of enlarged lymph nodes[4].

The current diagnostic gold standard for synovial sarcoma is demonstration of the t(X;18) (p11.2,q11.2) translocation using reverse transcriptase polymerase chain reaction (RT-PCR), involving fusion of the SYT (Synonyms: SS18-synovial sarcoma translocation, chromosome 18) gene on chromosome 18 to either the SSX1 (synovial sarcoma, X breakpoint 1) or the SSX2 (synovial sarcoma, X breakpoint 2) gene on chromosome Xp11. However, this method is limited in clinical practical work because of the cost, time, and availability of special equipment. While immunohistochemical staining is helpful in diagnosis, it unfortunately is not as accurate as molecular/genetic testing. This is largely owing to the low and differing sensitivities and specificities of the currently available immunohistochemical markers including Bcl-2, EMA, cytokeratins (CK), MIC -2, calponin. A recent study reported that transducin-like enhancer protein 1 (TLE1) demonstrated a 92% positive predictive value and 100% negative predictive value. This is significantly better than other currently used immunohistochemical markers, and thus may preclude the need for more costly and time-consuming molecular testing in some cases[7].

Currently, information on the treatment for head and neck synovial sarcoma is limited, and there is no ideal and standard therapeutic strategy. It has been reported that a local recurrence rate is up to 80% after incomplete surgical excision without adjuvant radiotherapy. A wide excision combined with postoperative radiotherapy is traditionally recommended to decrease the risk of local recurrence. Systemic adjuvant chemotherapy remains debatable, but it may play important roles in preventing or postponing the occurrence of distant metastases, especially in high-risk patients with tumors > 5 cm in size or with positive surgical margins. Due to the complex and vital anatomical structures in the head and neck region, a wide surgical excision is unlikely to perform without sacrificing to nearby structures. Thus, postoperative adjuvant chemoradiotherapy seems to be of more importance for synovial sarcoma of the head and neck than tumors in other locations[3].

CONCLUSION

Synovial sarcoma is a rare malignant tumor which treatment is essentially surgical resection with high rates of relapse. Although traditional treatment for hypopharynx tumors has been laryngectomy, it can be modified for sarcomas at this area. In these cases, most important are the border's microscopic control and lack of lymphatic metastasis. Radiation therapy with or without chemotherapy appears to be responsible for the increased survival in recent decades. It is important for the otolaryngologist and head and neck surgeon to be familiar with this aggressive tumor, which carries high

mortality and morbidity. The appropriate early diagnosis and treatment can improve the prognosis and survival of patients.

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