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Case Report

Synovial sarcoma masquerading as thyroglossal cyst carcinoma

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ABSTRACT

Synovial cell sarcoma is very rare in head and neck. It is usually seen in the extremities in close relation to the bursae and joints. In head and neck, most of the cases reported are in the retrohyoid, parapharyngeal and retropharyngeal areas. Reporting the case of a 18 year old man who presented with signs and symptoms of thyroglossal cyst. Radiological findings were suggestive of a haemorrhagic thyroglossal cyst or a cystic neoplasm. Frozen section revealed a malignant spindle cell neoplasm of mesenchymal origin. A Sistrunk operation was done. Histopathological and immunohistochemical analysis yielded a diagnosis of monophasic synovial sarcoma. Patient underwent intensity modulated radiotherapy according to tumor board decision. Follow up PET Scans were negative. We are reporting this case to highlight the rarity of the condition and the high index of suspicion required to diagnose such a synovial sarcoma in this area. Treatment should be aimed at complete surgical resection of the tumor and appropriate adjuvant radiotherapy.

Keywords: Synovial sarcoma, Monophasic, Thyroglossal cyst

INTRODUCTION

Synovial sarcoma (SS) is an uncommon, highly aggressive and distinct soft tissue neoplasm, predominantly affecting young people, accounting between 8 and 10% of all soft tissue malignancies. The tumor originates primarily in the extremities, in close relation to tendon sheaths, bursae and joint capsules. Synovial sarcoma of the head and neck region is quite rare. Jernstrom described the first documented report of head and neck synovial sarcoma in 1954 in a case involving the pharynx.[1] Since then fewer than 100 cases have been reported in the literature.[2] The most commonly involved cervical sites are the neck, the retro and the parapharyngeal spaces.[3] We report such a rare case of synovial sarcoma masquerading as thyroglossal cyst carcinoma in a young adolescent male.

CASE REPORT

Clinical history and physical examination findings

A 18-year-old male presented with swelling in the anterior aspect of neck for 8 months with rapid progression of size in the last 2 months. He did not have dysphagia or hoarseness of voice. Clinical examination revealed a swelling of size 2×2 cms in the infrahyoid region at the level of thyroid cartilage in the midline. The swelling moved with deglutition and on protrusion of tongue. Indirect Larynngoscopy showed mobile vocal cords. There were no palpable lymph nodes in the neck. Hence a clinical diagnosis of thyroglossal cyst was made.

Investigations

Complete hemogram was normal and TSH was mildly elevated. USG thyroid showed hypoechoic heterogenous

space occupying lesion in the midline which was avascular with no significant calcifications. FNAC of the swelling showed atypical cellular clusters suspicious of malignancy probably medullary carcinoma of thyroid. Tumour markers such as serum Calcitonin was 2.8 (<8.8 pg/ml) and serum thyroglobulin was 9.27 (3-42 ng/ml).

Radiographic findings

Contrast enhanced computerised tomography of the neck revealed normal thyroid lobes and an isodense lesion with peripheral enhancement with suggestion of enhancing internal septae and mural nodularity in the right infrahyoid neck region, suspicious of either hemorrhagic thyroglossal cyst or intracystic neoplasm (Figures 1 and 2). Tc99M thyroid scan showed heterogenous tracer distribution in thyroid with no demonstrable cold areas.

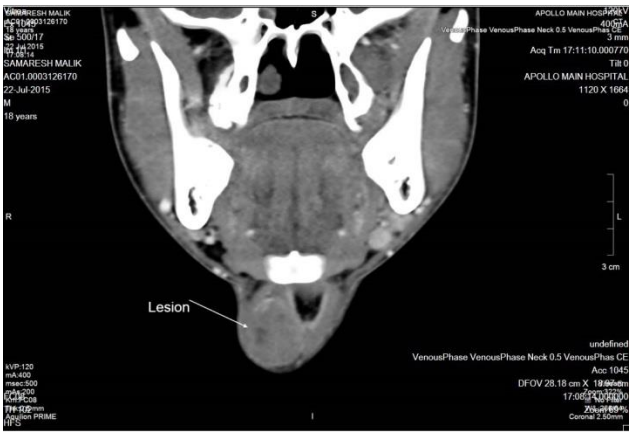

Figure 1: CECT Neck showing isodense lesion in the infra hyoid region on the right side.

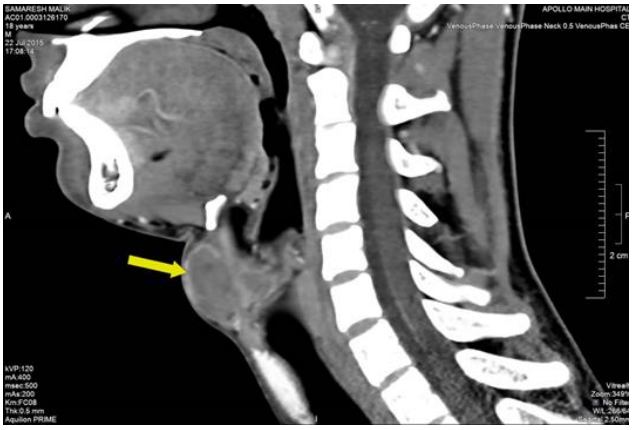

Figure 2: CECT Neck sagittal view showing the isodense lesion in the infra hyoid region with peripheral enhancement.

MANAGEMENT

Endocrinologist opinion was sought and planned for Sistrunk's operation. During surgery, a part of the lesion was excised and sent for frozen section that revealed a malignant spindle cell neoplasm, mesenchymal in origin. Hence the tumor along with the tract was dissected up to the hyoid bone and removed in toto along with the body of hyoid bone. The histological examination revealed a proliferation of spindle shaped cells arranged in sheets and fascicles (Figures 3 and 4).

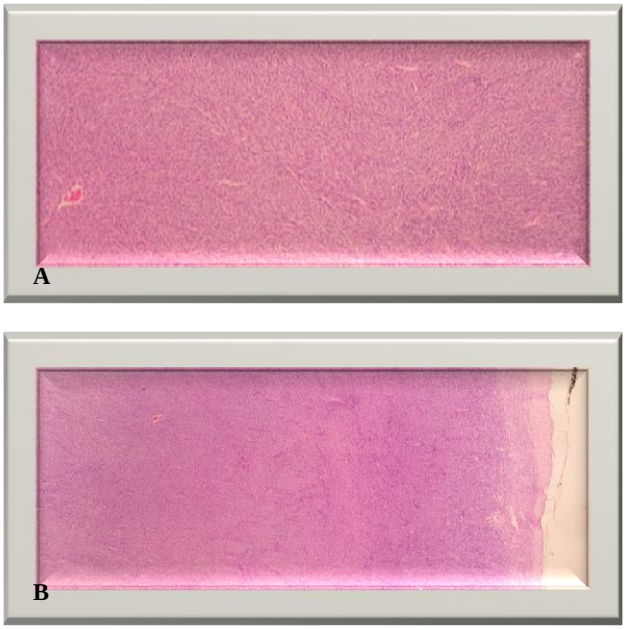

Figure 3 (A and B): Showing cellular neoplasm showing spindle cells in whorls, fascicles and interlacing bundles.

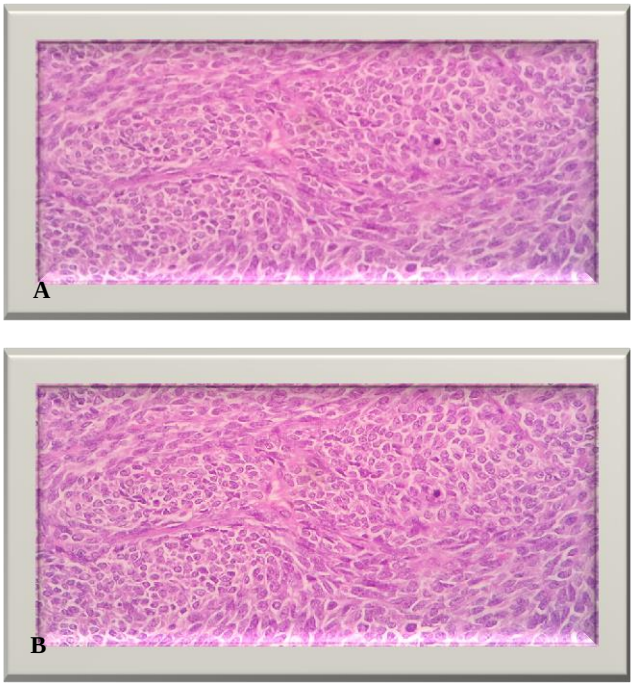

Figure 4 (A and B): Showing monomorphic spindle cells show occasional atypical mitosis.

On immunohistochemical analysis, tumor cells were positive for cytokeratin, vimentin, smooth muscle antigen, CD99 and epithelial membrane antigen, and negative for S-100, desmin and CD34. The histologic diagnosis of synovial sarcoma was made.

Two months after the surgery, medical oncologist opinon was sought and patient underwent 34 cycles of Intensity modulated radiotherapy (IMRT). PET CT scans done 2 months, 6 months and 12 months after IMRT were negative.

DISCUSSION

Synovial Sarcoma is a rare form of sarcoma in the head and neck region. In the past, monophasic synovial sarcomas have been reported in sites like lungs, nerves, gastrointestinal tract, liver, vulva and conjunctiva. Very few cases of Monophasic Synovial Sarcoma of thyroglossal cyst have been reported in the world literature.[3-6] It exists in children and young adults with preponderance in males.[4] This neoplasm usually follows a chronic protracted course with sudden exacerbation of symptoms occasionally.[6] The definitive diagnosis is attained only after a complete histopathological examination together with immunohistochemical evaluation of the excised tumor.[8] Synovial sarcoma usually presents as a slow growing, painless, well circumscribed solid mass. Although literature indicates the presence of some radiological findings including hemorrhage, calcification, heterogeneous intensity on T2 weighted images and heterogeneous enhancement pattern in this tumor, synovial sarcoma is frequently misdiagnosed as a benign entity owing to its smooth margins, cystic component and lack of aggressive infiltration.[8-10]

In our case, a young male presented with a cystic mass which was clinically and radiologically considered to be a benign cyst because of its chronicity, cystic nature and lack of aggressive infiltration.

Microscopically, synovial sarcoma consists of two predominant cell patterns which include the epithelial type and sarcoma like spindle cells. While biphasic type of the tumor consists of the distinct epithelial cells and spindle cells in different proportions, the monophasic type presents with a uniform pattern of a single type of cells. The poorly differentiated Synovial Sarcoma consists of predominantly epitheloid or round cell morphology and an increased mitotic activity. Immunohistochemical evaluation of tumor forms an indispensable tool for the definitive diagnosis of Synovial Sarcoma. Majority of synovial sarcoma exhibit immune positivity for epithelial membrane antigen and cytokeratins.[12]

The tumor cells in our case were positive for cytokeratin, vimentin, smooth muscle antigen, CD99 and epithelial membrane antigen, and negative for S-100, desmin and CD34.

The most accepted mode of treatment is surgical excision of the tumor mass with wide margins followed by postoperative radiation therapy especially in high risk cases. Obtaining adequate surgical margins is crucial for local tumour control. Chemotherapy has emerged as a promising treatment in the current times. Current literature points towards a multidisciplinary approach for treatment of the disease.[7]

The treatment of choice is surgery with or without radiotherapy and chemotherapy. Synovial Sarcomas commonly metastasize to lungs, skeleton and occasionally regional lymph nodes. Studies suggest that the use of adjuvant radiotherapy following surgery has a beneficial effect however neoadjuvant radiotherapy has yet to show any benefit.[13]

Survival rates were associated with tumor location, size, and extension. Treatment of synovial sarcoma of the head and neck should be directed towards complete surgical resection. Given the known sensitivity of synovial sarcoma to contemporary chemotherapy, a multimodality approach should be considered in the perioperative setting, especially in high risk patients.[14]

CONCLUSION

Monophasic Synovial sarcoma of the thyroglossal cyst are very rare. Because of rarity of this disease and a benign presentation, diagnosis of this entity presents as a challenge to surgeons, radiologists and pathologists. A high index of suspicion is required for this type of presentation especially in children and adolescents. An increased awareness of this disease with more representation of cases can assist in a definitive diagnosis and prompt treatment.

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