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Retropharyngeal synovial sarcoma—A rare location

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Abstract

Synovial sarcoma is a rare malignant tumor that derives from a mesenchymal precursor stem cell which is unrelated to mature synovial tissue. Synovial sarcoma in the head and neck region is quite rare accounting for only 6.8% of all sarcomas in clinical practice. We report on a case of low grade monophasic synovial sarcoma involving the retropharyngeal muscles in a 26-year-old female who presented with dysphagia and progressive dyspnoea.
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Keywords: Synovial sarcoma; Retropharyngeal muscles; Soft tissue sarcoma; Head and neck cancer

1. Introduction

Synovial sarcoma is a fairly common tumor usually involving the lower limbs of young adults; however, its localization in the head and neck region is quite rare accounting for only 6.8% of all sarcomas [1]. This report is of a 26-year-old female with low grade monophasic cell synovial sarcoma involving the retropharyngeal muscles, who presented with dysphagia and progressive dyspnoea.

2. Case report

A previously healthy 26-year-old female presented with 2 months history of progressive difficulty to breathe and dysphagia. She had a muffled voice with mild respiratory distress in supine position. Examination showed a retropharyngeal soft tissue mass causing almost complete narrowing of the airway. Flexible fibro optic laryngoscope done by the ENT experts showed the pharyngeal mass to have no involvement of the vocal cord.

Plain X-ray of the cervical spine showed a large soft tissue mass displacing the trachea anteriorly. There was anterior scalloping of the cervical vertebral bodies (Fig. 1). Contrasted computed tomography (CT) scan arranged immediately demonstrated a large well defined poorly enhancing retropharyngeal soft tissue tumor compromising the airways anteriorly (Fig. 2a). The mass extents from the nasopharynx to the hypophaynx causing erosion on the anterior aspect of C2 to C5 vertebral bodies (Fig. 2b). No surrounding inflammatory changes were seen. Despite the close proximity of the mass to the major neck vessels, no vessel involvement was observed. Both the neural foramens were not widened, however possible extension of the tumor into the spinal canal cannot be excluded. Multiple small cervical lymph nodes were noted. In view of the chronic and benign appearance of the tumor, a preliminary radiological diagnosis of nerve related tumor such as schwannoma or leiomyoma was made. MRI was then suggested to exclude extension into the spinal canal.

In anticipation to complete obstruction of the oropharynx, an emergency tracheostomy was performed under local anaesthesia on admission. MRI performed two weeks later showed a large retropharyngeal soft tissue mass which does not extend into the spinal canal. The tumor was predominantly of low signal in T1-weighted images, high on T2-weighted images and demonstrates heterogeneous enhancement. There was infiltration of the mass into the left anterolateral aspect of the vertebral

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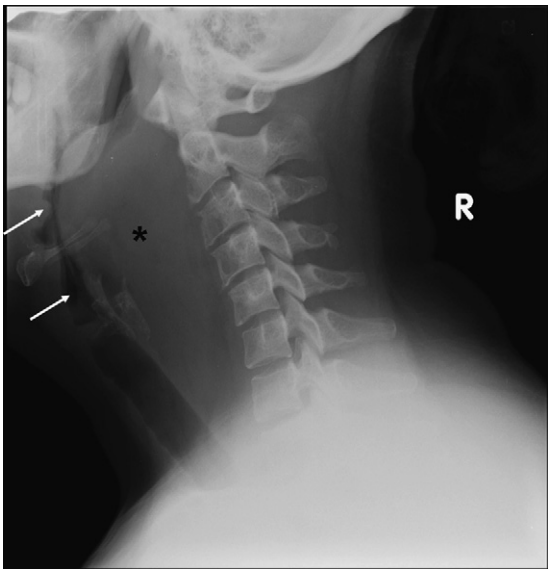

Fig. 1. Plain lateral radiograph of the neck in soft tissue window shows a large retropharyngeal soft tissue mass (asterisk) causing narrowing and anterior displacement of the trachea (white arrows). There was anterior scalloping of the cervical vertebral bodies.

body. High signal changes were also noted in the adjacent left stenocledomastoid muscle suggestive of tumor infiltration (Fig. 3).

A CT guided biopsy was performed under general anaesthesia as patient was unable to lie down. Histology showed proliferation of spindle cells in fascicles that were immunoreactive to vimentin and actin spindle favoring the diagnosis of low grade synovial sarcoma (Fig. 4). No obvious areas of necrosis noted. Patient was referred to oncology clinic for further management, however; she had opted for traditional therapy and was lost to follow-up.

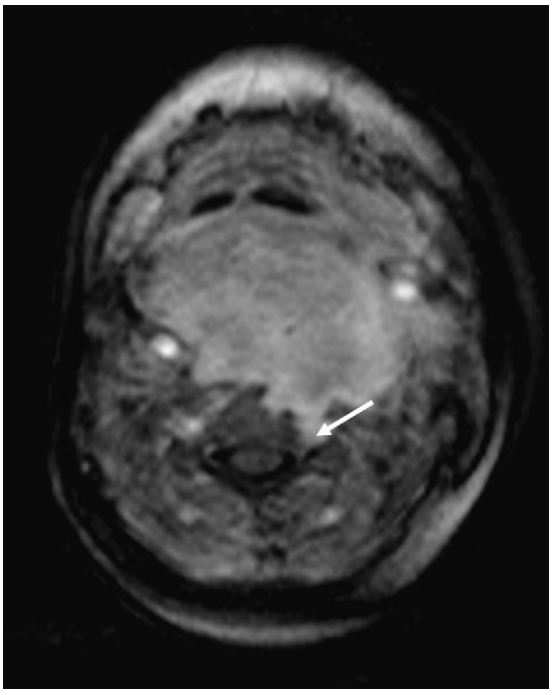

Fig. 3. Contrasted axial MRI of the neck shows the mass enhances heterogeneously with no evidence of extension into the spinal canal (arrow). There is infiltration of the mass into the left anterolateral aspect of the vertebral body. High signal changes were also noted in the adjacent left stenocledomastoid muscle.

3. Discussion

Pharyngeal tumors are rare accounting for 0.5% of all head and neck neoplasms. Tumors at these sites have a wide variety of histology, majority of these being benign. In a case series of pharyngeal tumors, 65.4% of the tumors were found to be benign while the rest were malignant. Of these, salivary

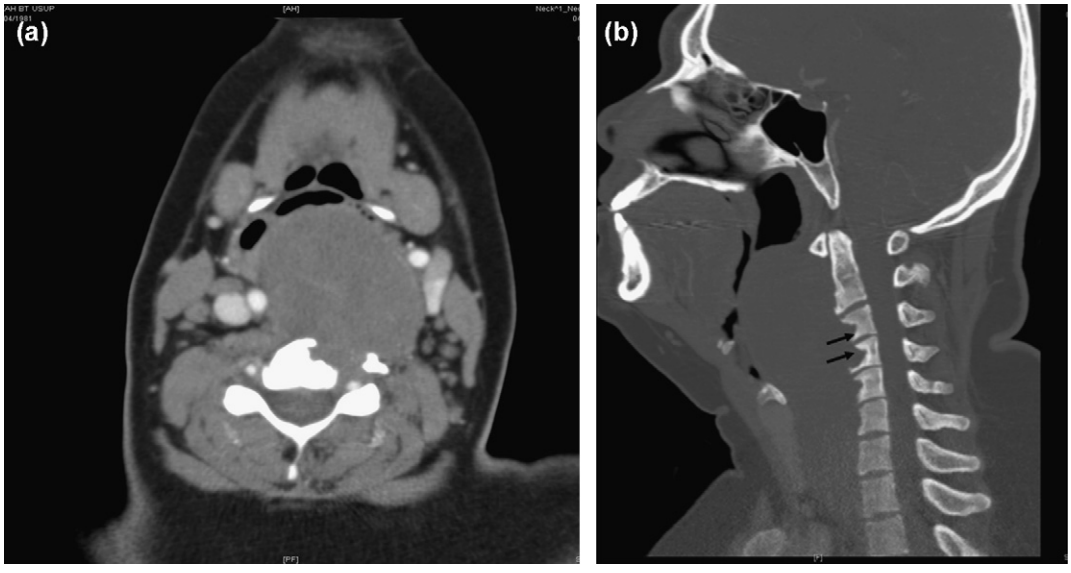

Fig. 2. CT in (a) axial soft tissue and (b) sagittal bone window show a well demarcated and homogeneous retropharyngeal mass more towards the left with smooth margins causing narrowing and displacement of the larynx and pharynx. There is associated bony erosion of the anterior vertebral bodies (arrows). The neural foramen is not widened. Small cervical lymph node noted.

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