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Cervical Pleomorphic Liposarcoma: A Rare Case Report and Role of Radiology

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ABSTRACT

Background: Liposarcoma is a rare type of adult soft tissue sarcoma with majority of them arising in the thigh and retroperitoneum. Liposarcomas are extremely rare in the head and neck region. Usually asymptomatic, they may cause discomfort, be painful or even life-threatening especially when voluminous and located in the upper aerodigestive tract. We report a rare case of a cervical pleomorphic liposarcoma, to illustrate the imaging and pathologic findings.

Case presentation: A 70 year old male presented with right sided neck swelling with associated weight loss and loss of appetite. Physical examination showed a firm lump in the right posterior neck region. Computed topography (CT) scan revealed a well-circumscribed heterogenous mass with enhancing solid component in the right posterior cervical space (PCS) for which the patient underwent exploration and complete excision with good results.

Discussion and conclusion: Head and neck liposarcomas are extremely rare entities that might be life-threatening. Although clinical and imaging manifestations are not specific, imaging helps in pre-operative planning and anatomical localisation. Pathological examination is crucial to confirm the diagnosis.

Keywords: Pleomorphic, Liposarcoma, cervical

Background

Although soft tissue sarcomas as a group are not uncommon diagnoses, liposarcomas are much rarer and encompass a wide range of diagnoses. Liposarcomas are extremely rare in the cervical region. Liposarcomas of the head and neck are usually early stage, low grade, and with fewer nodal metastases than conventional liposarcomas. We present a case of pleomorphic liposarcoma involving the right posterior cervical space diagnosed in a 70 year old male who presented with a right sided neck mass, for which he underwent neck exploration and complete excision with good results.

Case presentation

A 70-year-old male with history of smoking and hypothyroidism was referred to the surgical outpatient clinic with chief complaint of a swelling on the right side of the neck (*fig.1*), which he had noted for the past 3 years with increase in size over the last 2 months. There was associated history of loss of appetite and significant weight loss. There was no dyspnoea, dysphagia, dysphonia, cough, or expectorations. Physical examination showed a well-circumscribed swelling in the right side of the neck. The swelling was non tender, non-mobile and firm in consistency.

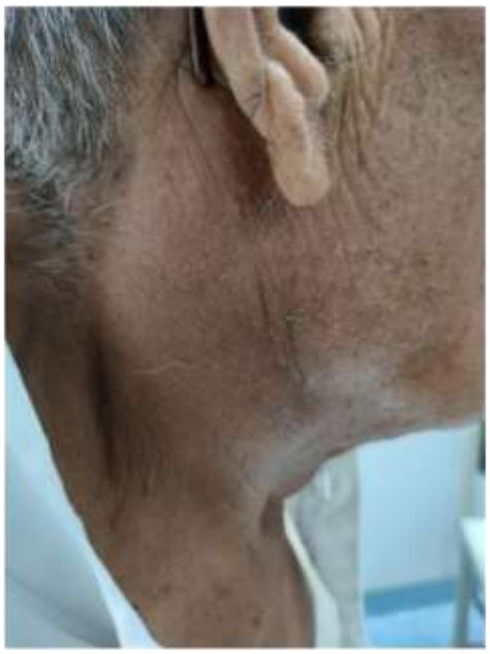

Figure. 1 Photograph showing a well circumscribed swelling in the right cervical region, which was firm in consistency on palpation.

Ultrasonography revealed a well-defined heterogenous echogenicity lesion in the right posterior neck space with multiple internal cystic areas and echogenic components. Contrast enhanced CT showed a solitary well defined ovoid heterogeneously enhancing mass (*fig. 2A-B*) in the right PCS displacing the overlying sternocleidomastoid muscle laterally, with intact fat planes. Internal jugular vein (IJV) and carotid artery were displaced anteromedially with focal narrowing of IJV at places *(fig. 3A-D).* A differential of schwannoma of PCS probably originating from cranial nerve XI was considered. Fine needle aspiration cytology (FNAC) was done which raised suspicion of malignant cells.

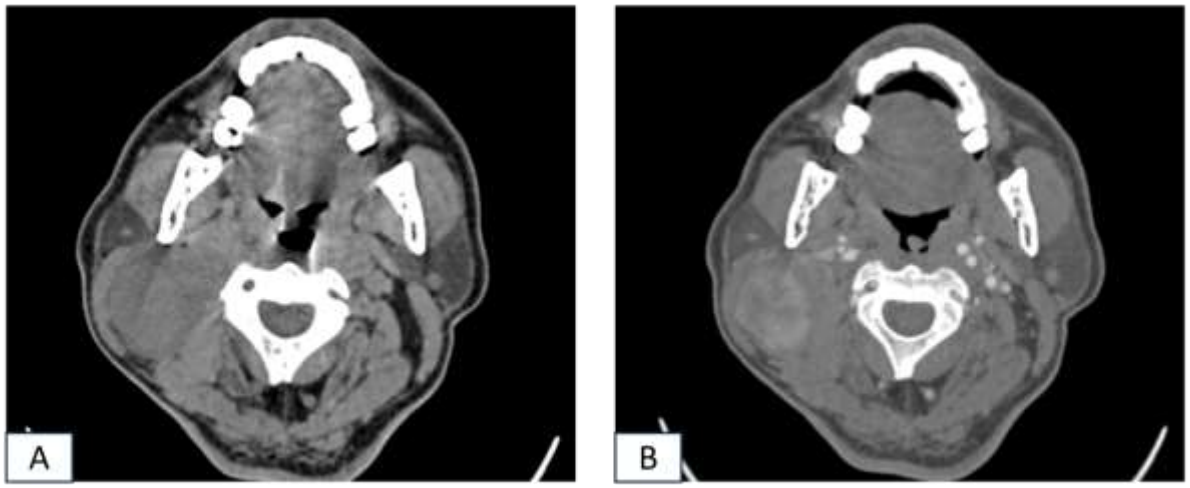

Figure 2.A & B: Axial non contrast (A) and contrast enhanced CT (B) of the neck at C2 vertebral level showing well defined heterogenous attenuation soft tissue mass in right posterior cervical space.

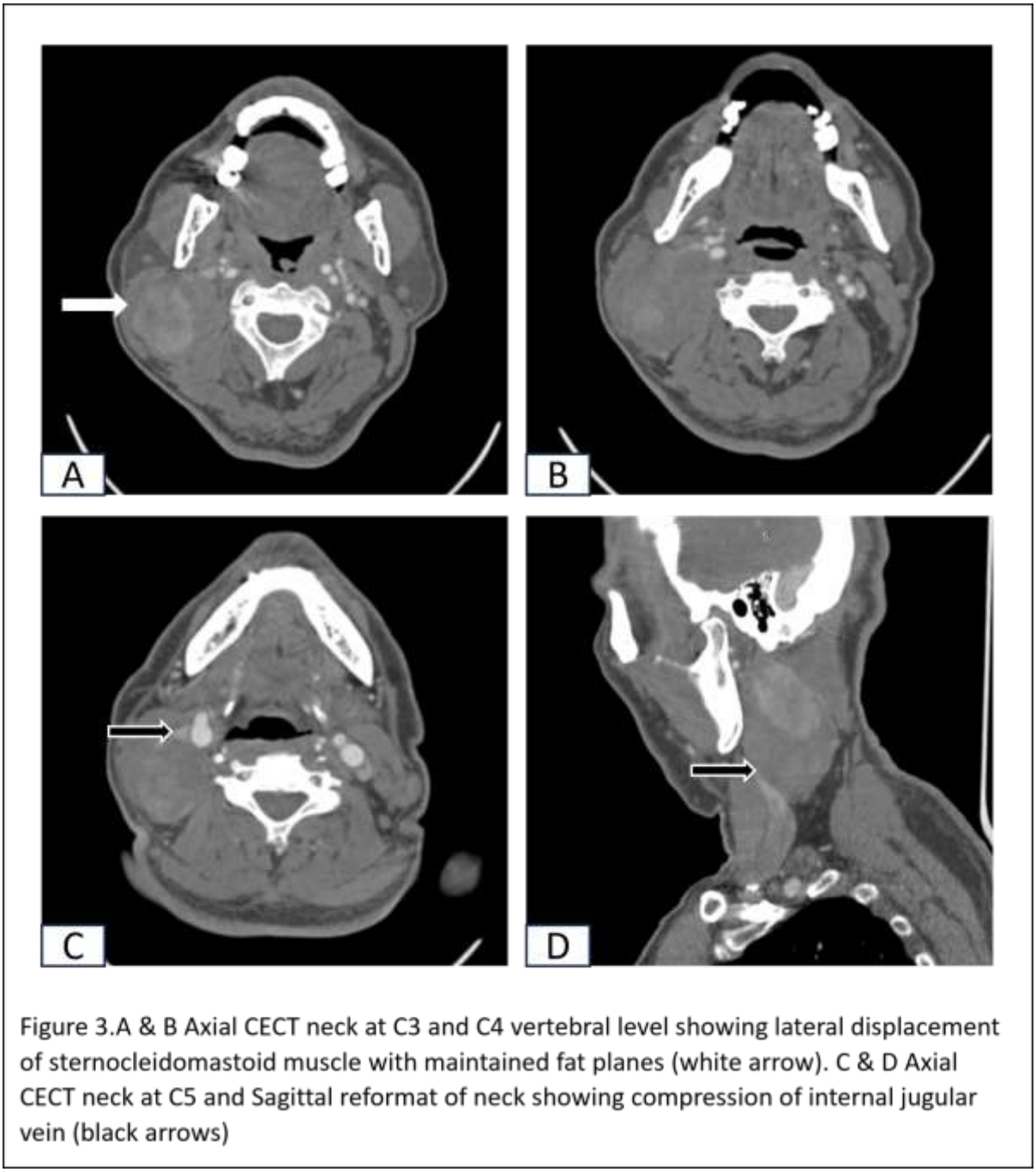

Figure 3.A & B Axial CECT neck at C3 and C4 vertebral level showing lateral displacement of sternocleidomastoid muscle with maintained fat planes (white arrow). C & D Axial CECT neck at C5 and Sagittal reformat of neck showing compression of internal jugular vein (black arrows)

Surgical exploration and excision of the mass was planned using CECT. A well circumscribed ovoid mass was excised which had free margins with carotid space vessels and was not adherent to the adjacent structures. The postoperative course was uneventful. Histopathology examination (*Fig. 4*) showed a ovoid mass with nodular surface with yellow tan ill-defined multilobular areas in cut section. Microscopy revealed presence of high-grade pleomorphic lipoblasts with cytoplasmic vacuolation and scalloped nuclei. A diagnosis of pleomorphic liposarcoma was finally made according to the FNCLCC system.

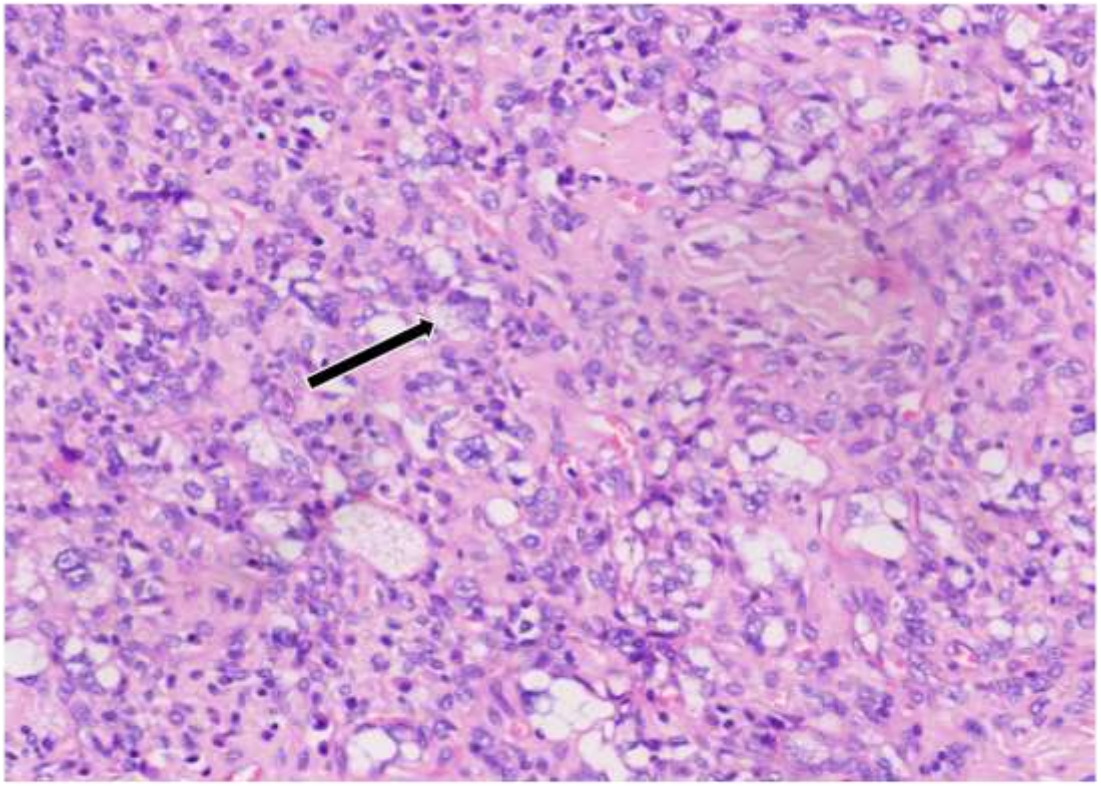

Figure. 4 shows tumour cells arranged in sheets. Cells shows high grade pleomorphism with multinucleation. Pleomorphic lipoblasts with cytoplasmic vacuolation and scalloped nuclei are noted (*black arrow*).

Discussion:

Liposarcoma occurs most frequently in patients in their seventh decade. Liposarcoma is a rare mesenchymal tumour with thigh, buttocks and retroperitoneum being the most common sites. Liposarcoma accounts for approximately 15% of all soft tissue neoplasms with their prevalence in the head and neck region representing only 2–9% of all malignant mesenchymal tumours [1,2].

Radiologically, the cervical liposarcomas are commonly encountered in the posterior cervical space [3]. As they are extremely rare and imaging features are nonspecific, diagnosis is usually made after histopathological assessment. CT appearances of lipomatous tumours are based on the distribution of the fat and non-fat components. Features which may suggest a liposarcoma include fatty mass with irregularly thickened enhancing linear and nodular solid components [4]. However, in our case no macroscopic fat was seen in the lesion. Schwannoma of PCS remains a close differential as it is more common in this anatomical location. Other differential diagnosis in the region includes lymphatic and vascular malformations, reactive or sarcoid adenopathy, benign tumours like lipoma, schwannoma or neurofibroma, malignant tumours like non-Hodgkin lymphoma and squamous cell carcinoma [5].

Histologically, there are 5 histological subtypes: well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated. PLS is very rare, and accounts for only 5% to 10% of lipomatous tumours [6]. High grade liposarcomas (PLS and dedifferentiated subtype) generally do not show substantial amount of fat or other specific radiological features, hence are inseparable from other sarcomas [4].

Previously, Dhruv Gupta et al described a cervical liposarcoma which showed heterogenous enhancing mass lesion with an irregularly enhancing wall in right side of the neck superficial to sternocleidomastoid with stranding of subcutaneous fat planes [7]. Y. Eang et al described a case of cervical liposarcoma which presented as a large neck mass with a lipomatous region with thick septa, a low-attenuation high-water-content component, and a heterogeneous high attenuation calcified area with no significant post contrast enhancement [8]. However, in our case no enhancing wall, fat stranding or calcification was seen.

Typical features of cervical liposarcoma according to literature review include fat containing lesions with solid and cystic components, the solid component showing post contrast heterogenous enhancement. Our case does not match the typical description in view of its fat deficient nature. The lesion was also non-adherent to adjacent structures. Imaging features being non-specific, tissue biopsy confirmed the diagnosis. Imaging is however invaluable for pre-op planning and assessing the relation to adjacent structures especially the vessels.

Conclusion:

PLS of the cervical region is extremely rare, with the posterior cervical space being the most common location in the neck. Although, the imaging features are nonspecific, contrast enhanced CT plays a crucial role in lesion characterization, anatomical localization, ruling out other aetiologies and pre operative planning. More studies are required to develop more reliable imaging features to suggest the diagnosis of cervical liposarcoma.

List of abbreviations:

CT: Computed topography

FNAC: Fine needle aspiration cytology

FNCLCC: Federation nationale des centres de lute contre le cancer

PLS: Pleomorphic liposarcoma

PCS: Posterior cervical space

Declarations:

Ethics approval and consent to participate: The consent for this report was obtained from the Institutional Ethics Committee and Informed Consent was taken from the patient.

Consent for publication: An informed consent for publication purposes was obtained from the patient.

Availability of data and materials: The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interests: The authors declare that they have no competing interests.

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Authors' contributions: Author 1 was involved in the literature review, formal analysis, diagnosis and writing original draft. Author 2 was involved in the diagnosis, validation, supervision, writing-reviewing and editing. Author 3 was involved in diagnosis and reviewed the manuscript for insightful remarks. Author 4 was involved in the surgery procedure and reviewed the manuscript. Author 5 was involved in the pathology study and reviewed the manuscript. All the authors have read and approved the final manuscript.

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