

## Primary Leiomyosarcoma of Internal Jugular Vein: A Rare Entity

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**Abstract:** Vascular leiomyosarcoma is a rare tumor arising from smooth muscle cells in tunica media of venous walls. It involves inferior vena cava predominantly; involvement of jugular veins is extremely rare. This case report is of a middle aged lady who was diagnosed as leiomyosarcoma of internal jugular vein (IJV). She was treated by segmental resection of the vein followed by adjuvant External beam radiation therapy (EBRT) and ifosfamide based chemotherapy. Leiomyosarcoma can be considered as a rare differential diagnosis in patients presenting with lateral neck mass.

**Keywords:** Leiomyosarcoma, Internal Jugular Vein, External Beam radiation Therapy.

### INTRODUCTION

Soft tissue sarcomas are rare malignant tumors of mesenchymal origin encompassing 0.7 % of all cancers and leiomyosarcoma accounts for only 5-10 % of them [1]. Majority of Leiomyosarcoma originate from four sites namely intraabdominal/Retroperitoneal, cutaneous, subcutaneous and vascular. Three fourth cases of intravascular leiomyosarcoma affect retroperitoneum; predominantly inferior vena cava [2]. A few hundred cases have been reported since Perl's first description in 1871 [3]. Vascular Leiomyosarcomas involve veins more often than arteries and represent less than 2% of all leiomyosarcomas [4, 5]. Vascular Leiomyosarcomas rarely involve extracaval venous branches usually the venous branches of the lower extremity [3]. In this report, we describe case of a 45-year old female patient with a primary Leiomyosarcoma of the IJV.

### CASE REPORT

A 45 year old nulliparous female resident of Kolar district of Karnataka reported to the Department of Radiotherapy, Victoria Hospital, Bangalore with complaint of painless neck swelling on right side since last 3 months. The swelling was progressively increasing in size to reach the present size (4x3 cm). Earlier patient was evaluated at other hospital with Ultrasound examination which revealed a large lobulated, heterogeneously hypodense mass lesion noted in right side of neck measuring 4.5x3.6x2.6 cms

causing compression of right IJV-probably a *mesenchymal* tumor. Fine needle aspiration cytology (FNAC) suggested a diagnosis of spindle cell variant of paraganglioma. She underwent total abdominal hysterectomy three years back for multiple fibroid uterus.

On examination a firm oval mass was noted just anterior to anterior border of sternocleidomastoid. The mass had restricted mobility in all directions and measured 4.5 x4 cm. Two palpable lymph nodes of sub centimeter size were noted in right level IA, II lymph nodal region.

She was evaluated for any primary tumor in upper aero-digestive tract by upper GI endoscopy & bronchoscopy which were normal. Contrast enhanced CT scan of neck and thorax revealed a lobulated fairly defined soft tissue attenuating lesion measuring 7x2.9x4.8 cm with heterogeneous predominantly peripheral contrast enhancement probably neoplastic. Lesion was extending from C7 to T3 vertebral level. Anteriorly it was closely abutting sternocleidomastoid and invading IJV. Medially the lesion was abutting lateral margin of right lobe of thyroid. Few enlarged submental, supraclavicular, submandibular & upper jugular lymph nodes were also noted, largest being right upper jugular measuring 12x10 mm.

Patient was referred to surgical oncologist, underwent right radical neck dissection and post-op histopathology report showed spindle cell sarcoma arising from tunica media of IJV, infiltrating full thickness of vessel wall and soft tissue laterally. Neoplastic cells showed moderate pleomorphism & mitotic count of 10-25/10 high power fields (HPF). Soft tissue cut margin was focally positive for tumor cells. All eleven lymph nodes were found reactive. Further immunohistochemistry (IHC) test showed cells positive for SMA, desmin and negative for cytokeratin (CK),

CD-34 and S-100. Ki index was 50%. Hence a final diagnosis of Leiomyosarcoma of right IJV was made. In view of focally positive margin and lateral soft tissue involvement by tumor cells, patient received adjuvant EBRT to the right side of neck using Antero-posterior and postero-anterior (AP-PA) portals to a total dose of 50 Gy in 25 fractions over 35 days with RTOG grade-I skin toxicity and grade-II dysphagia. Further she received 6 cycles of ifosfamide & Adriamycin based adjuvant chemotherapy. Presently patient is on routine 3 monthly follow up & disease free since 1 year.

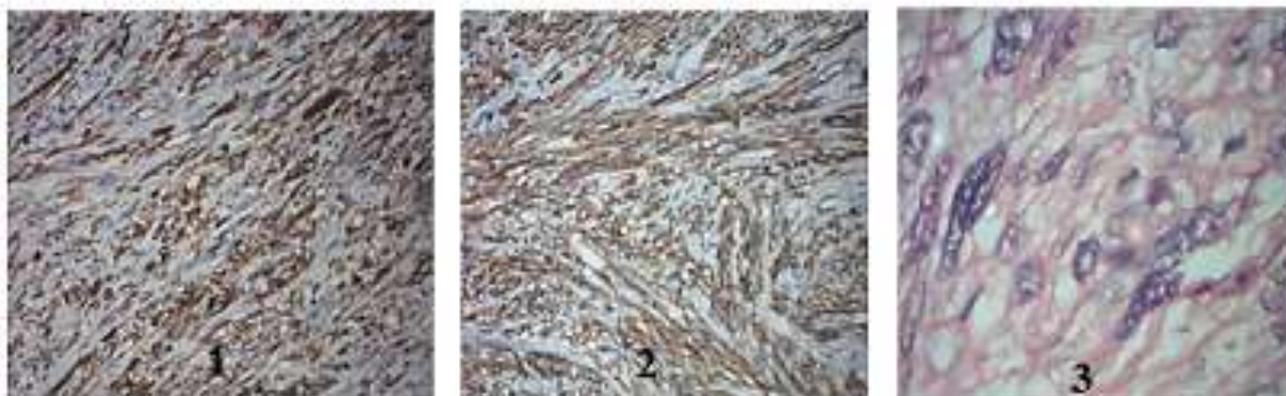


Fig. 1: tumor cells positive for desmin; Fig. 2: tumor cells positive for SMA; Fig. 3: HP picture of affected IJV

## DISCUSSION

Vascular leiomyosarcoma presenting a small proportion of soft tissue leiomyosarcoma [2] derived from smooth muscle of venous vessel walls [3], but a single case of arterial origin has also been reported. 75% of the cases involve IVC [6] and leiomyosarcoma involving smaller vessels are rare. These tumors usually lead to nervous or arterial compression due to increased pressure within the neurovascular sheaths [2] and they often protrude through small lumina of adjacent venous branches [7]. In this patient, the tumor was localized in the right IJV which is a very rare site of involvement. To our knowledge only few such cases have been reported in literature [8], wherein the tumor involved both internal external jugular vein and subclavian vein. The case is unique as the tumor involved internal jugular vein alone. In the current literature unusual manifestations of intravascular leiomyosarcoma were described for venous branches of the lower extremity [9], whereas only few case reports of tumor manifestation of the upper extremity, the head and neck region and azygos vein were found [3, 10-12].

## Differential diagnosis

Mesenchymal tumors, myofibromatosis, Fibromatosis, myofibroblastic sarcoma, synovial sarcoma, fibrosarcoma and NOS (not otherwise specified) sarcoma can be considered [13]. Complete surgical resection of the vessel segment is the therapy of choice. Leiomyosarcomas of a vascular origin appear to be associated with a more aggressive tumor growth

and poorer prognosis compared to respective tumors of the soft tissue [9]. Incomplete tumor resection or margin positivity requires adjuvant radiation therapy. Tumor size and localization are of prognostic value [11]. The intravascular growth of the sarcoma predisposes for hematogenous metastases [14]. Hence pulmonary metastasis has to be considered in the oncological follow up.

## CONCLUSION

In this report, we present a very rare case of intravascular leiomyosarcoma involving internal jugular vein in isolation. Such a diagnosis requires extreme degree of careful clinical examination, high accuracy of imaging studies. Complete tumor resection of the tumor with adequate margins is the primary treatment modality; however such approach may not be always possible due to difficulties associated with achieving clear resection margins, hence adjuvant radiation and chemotherapy also make an important part of treatment.

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