# Proceeding Book

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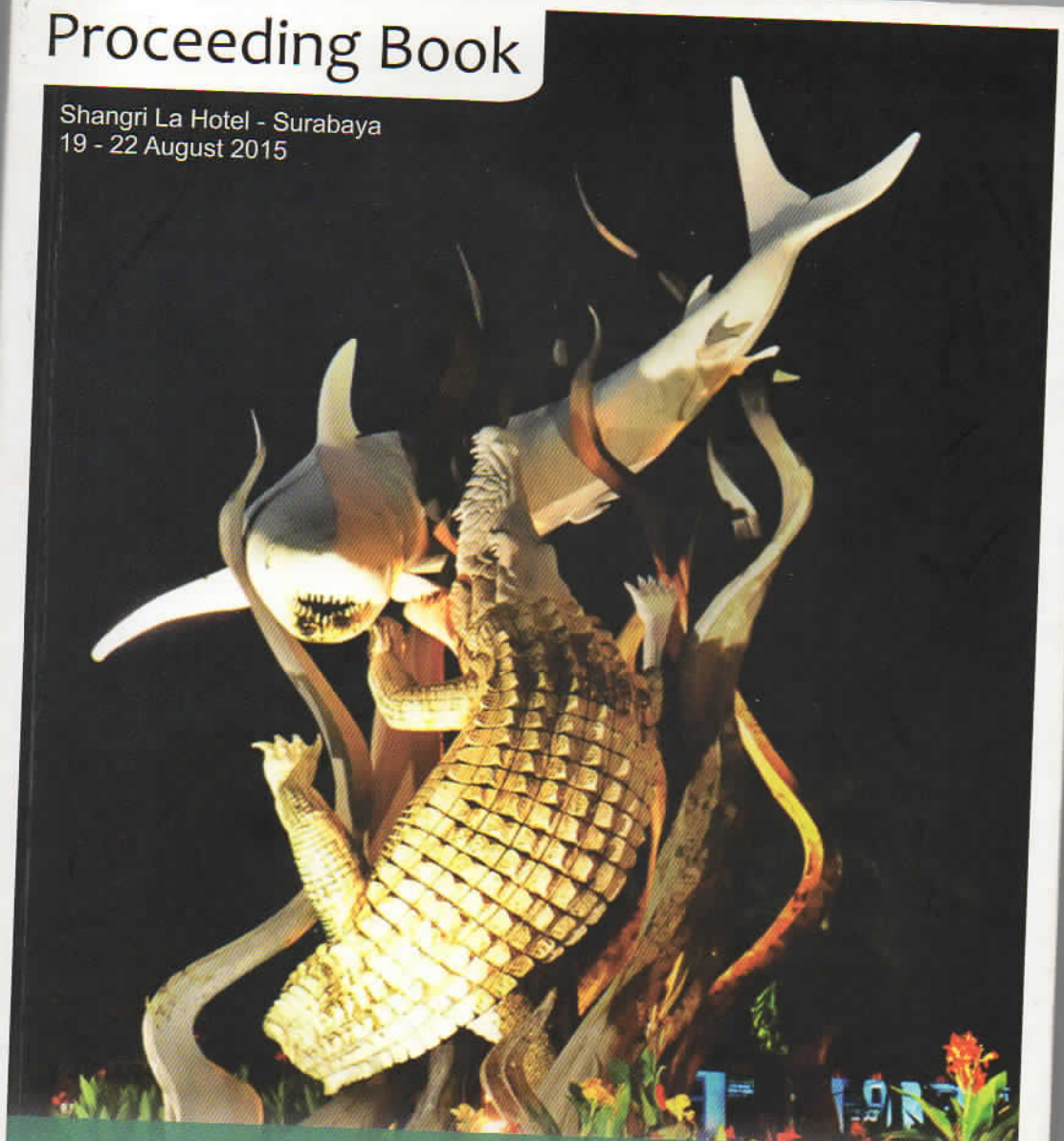

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# Head and Neck Leiomyosarcoma: A Case Report

**Hermawan, N.G.D** [1], Golden, N [2]
[1] *Medical Faculty,Udayana University,Bali,Indonesia*
[2] *Neurosurgery Subdivision,Sanglah General Hospital,Bali,Indonesia*

**Introduction**
Leiomyosarcoma is malignant tumor of smooth muscle which accounts for only 4% of head and neck sarcomas. The tumor is commonly encountered as a slow growing, discrete firm, and non-ulcerated painless mass. The physical appearance of these tumors can be deceptively benign and can be mistaken for non-malignant conditions. The most common metastatic sites are lung, liver, and peritoneal cavity whereas bone and brain metastases are rare. An early diagnosis and aggressive initial treatment remains the mainstay of therapy for a good prognosis

**Case Description**
A-56 year old woman presented with persistent headache for about 3 weeks and it was not getting well with many medicines. She also noted a progressive bulging mass over her left neck until occipital head region. Physical examination revealed a large fixed hard mass, about 10 cm in diameter and located at her right posterior cervical to occipital region. MRI of head and neck region showed lobulated mass, size3.35 x 4.95x4.47 cm on clivus-CV-C2 Left side (Carotid space) with strong homogenous enhancement after contrast administration. This tumor extended to left side cerebellum, left retroparyngeal space, insisted on and narrowing pharynx, internal carotid artery and to the anterior and left side spinal cord. Open biopsy was performed with the pathological result was Leiomyosarcoma. Surgical tumor resection was then done. At the same time cervico-occipital fixation was applied. After operation, headache was completely recovered. She was put on radiotherapy. In the follow-up the local tumor was well controlled.

**Key words**: Head and neck Leiomyosarcoma, surgical resection, radiation treatment

*Contact Author :*
*Full Name :Ngakan Gede Dwija Hermawan*
*Address : Jln.Batuyang Gang Pinguin No.9A Batubulan Gianyar No.9A Batubulan,Gianyar,Bali*
*Phone : 081239612185*
*Email :Ngagedwi_hermawan@yahoo.co.id*

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## INTRODUCTION

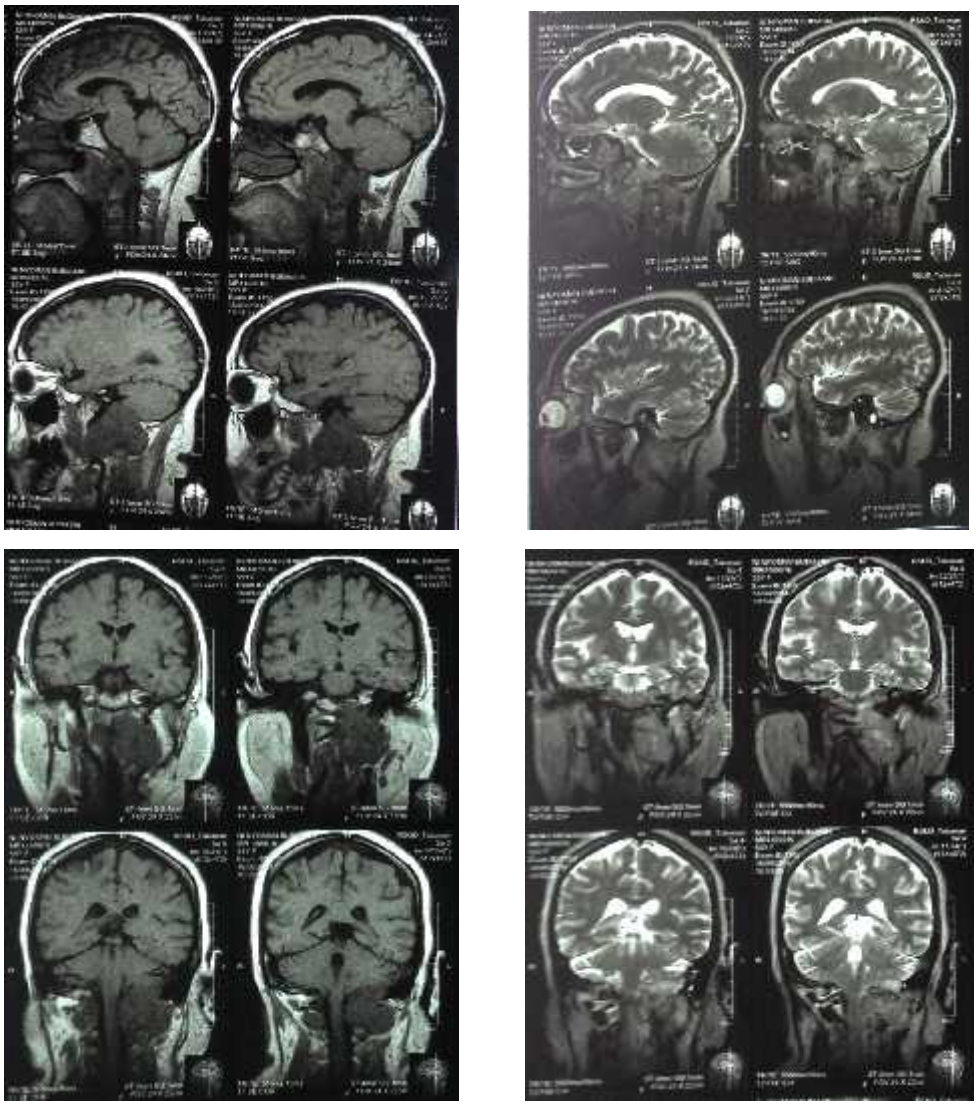
Head and neck neoplasms of mesenchymal origin are uncommon The majority of these tumors represent metastatic disease from other primary sites.[1] Leiomyosarcoma is an uncommon malignant neoplasm of smooth muscle origin, usually occurring in the retroperitoneum, subcutaneous tissues, blood vessels,gastrointestinal tract, genitourinary tract, and the uterus.[1,2] The tumor tends to invade and spread locally, but could also have an aggressive growth pattern with hematologic dissemination. The most common metastatic sites are lung,liver, and peritoneal cavity, whereas bone and brain metastases are rare. Soft-tissue sarcomas are rare in the head and neck, accounting for 1% of malignant neoplasms in this area.Leiomyosarcoma (LMS), a sarcoma derived from smooth muscle that can be either cutaneous (derived from errectorpili muscles of hair follicles) or subcutaneous (derived from vascular smooth muscle), represents 1% to 4% of such tumors.[2,3] As such, LMS of the head and neck is an extremely rare entity that because of its infrequency has been associated with both delayed diagnosis and misdiagnosis. Immunohistochemical stains can help distinguishing these tumors from meningioma.Postoperative radiation and chemotherapy should be considered.[3,4,5]

We describe the course of a patient with a head and neck leiomyosarcoma and review the literature.

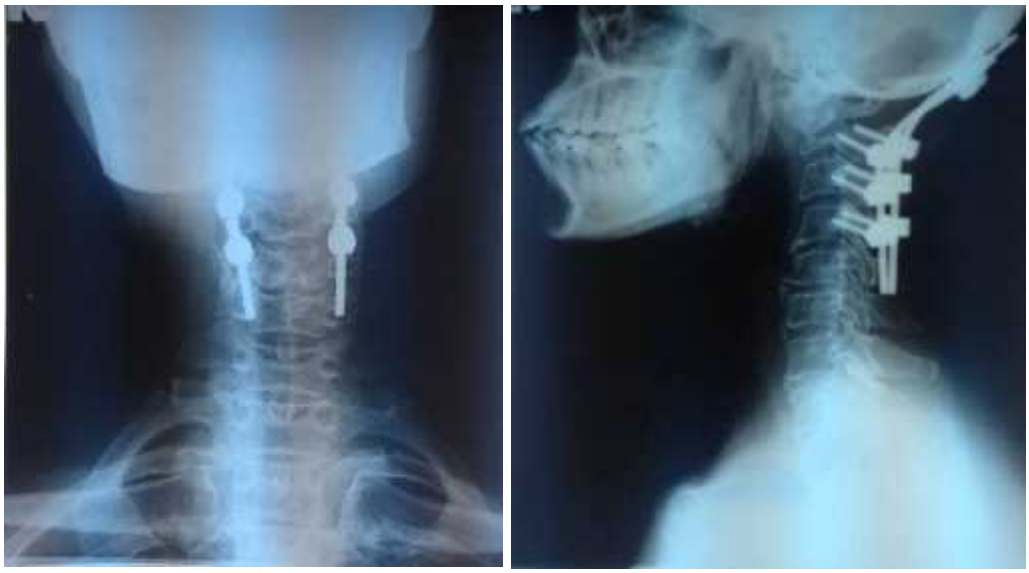
## CASE DESCRIPTION

A-56 years old woman presented in November 2014 with chief complaint persistent headache for about 3 weeks and it was not getting well with many medicines. She also noted a progressive bulging mass over her left neck until occipital head region. On history taking revealed in September 2003, she underwent gynecological surgery with the pathological report was Leiomyosarcoma. After surgery she was put on chemotherapy treatment. Since then the local tumor seemed to be well controlled. On physical examination revealed a large fixed hard mass, which was about 10 cm in diameter and located at her right posterior cervical to occipital region. There was no hemodynamic problems. Neurological examination demonstrated clear consciousness.The muscle power of her extremities was grade 5. MRI of head and k region showed lobulated mass, size 3.35 x 4.95x 4.47 cm on clivus-CV-C2 Left side (Carotid space), strong homogenous enhancement after contrast administration. This tumor extended to left side cerebellum, left retroparyngeal space, insisted on and narrowing pharynx, internal carotid artery and to the anterior and left side spinal cord (Figure 1).

Open biopsy performed in December 2014 with the pathological result was Leiomyosarcoma. Surgical tumor resection was then done on January 13$^{th}$2015. At the same time cervico-occipital fixation was applied (Figure 2). After Operation, headache was completely recovered. She was put on radiotherapy in April 2015. In the follow-up the local tumor was well controlled.



(Figure 1 : MRI )



(Figure 2: Cervico-occipital fixation )

**DISCUSSION**

Leiomyosarcoma is the malignant of smooth muscle tumors. Soft tissue sarcomas of the head and neck are rare tumors.[1] Smooth muscle is derived from primitive mesenchyme and is found mainly in blood vessels, erector pile musculature of skin, circumvallate papilla, primitive mesenchyme and myoepithelial cells of salivary glands.[1,2] The diagnosis of leiomyosarcoma is confirmed by ultrastructural features of smooth muscle cells and immunohistochemistry.[2,3,4] The tumor cells are elongated with tapering cytoplasmic processes with elongated, convoluted nuclei,pinocytic vesicles, and basement membrane material around the cytoplasmic membrane.[4,5] The differential diagnoses, which include malignant astrocytoma, malignant fibrous histocytoma, and meningioma, were excluded by immunohistochemical testing.[6] Aberrant mesenchymal differentiation and metastasis are the other possible modes of origin. The usual route of metastatic spread for leiomyosarcoma is theorized to be the bloodstream to the lungs.[5] Head and neck metastases are usually seen in the advanced stage of disease. The clinical features of skull head and neck metastases include local swelling, local pain, and neurological deficits, according to on the metastatic sites. The differential diagnosis of this rare tumor may be problematic. The physical appearance can be deceptively benign and may be mistaken for non-malignant conditions. Therefore, the diagnosis was supported by immunohistochemical and ultrastructural investigations. Histologically, the tumor is characterized by prominent interlacing bundles and fascicles of elongated ‘‘cigar-shaped’’ bluntended nuclei, prominent nucleoli and abundant eosinophilic cytoplasm spindle cells.[2,3,5]

Surgical resection is the principal treatment method for the soft tissue sarcoma. Surgery cannot alter the course of the underlying disease but can relieve the local discomfort and mitigate the neurological deficits if present.[5] Because adjacent pseudocapsule is commonly infiltrated by the tumor cells and satellite lesions are often found at some distance from the main lesion, the margin of the excision should be at least 1 cm in all directions. Because of the proximity of adjacent neurovascular structures or vertebral column, en bloc resection and achieving these margins at all tumor planes is almost impossible in the head and neck region. Metastatic spinal leiomyosarcomas tend to symptomatically involve only one spinal level at the time of diagnosis and are known to recur locally.[7] Neck dissection is usually not essential because of rarity of metastatic lymph node.[2,4,5]

Radiation therapy may be used as the primary modality for palliation in certain patients with metastatic disease, mostly patients with bony metastases. In this scenario, radiation is recommended for symptomatic metastases only. 240 However, lytic metastases in weight-bearing bones such as the femur, tibia, or humerus also are considered for irradiation. Another scenario in which radiation might be appropriate is spinal cord compression due to metastases to the vertebral body extending posteriorly to the spinal canal. The goal of adjuvant radiation therapy is to decrease local-regional recurrence rates. Adjuvant radiation therapy can be given before surgery, after surgery, or in selected cases, during surgery. Preoperative radiation therapy has several advantages. It may minimize seeding of the tumor during surgery and it allows for smaller treatment fields because the operative bed has not been contaminated with tumor cells. Finally, radiation therapy for inoperable tumors may achieve adequate reduction to make them operable. The disadvantages of preoperative therapy are an increased risk of postoperative wound healing problems and the difficulty in planning subsequent

radiation therapy in patients who have positive surgical margins. If radiation therapy is given postoperatively, it is usually given 3 to 4 weeks after surgery to allow for wound healing. The advantage of postoperative radiation therapy is that the surgical specimen can be evaluated histologically and radiation therapy can be reserved for patients who are most likely to benefit from it. Further, the radiation therapy can be modified on the basis of margin status. The disadvantages of postoperative radiation therapy are that the volume of normal tissue requiring irradiation may be larger owing to surgical contamination of the tissue planes and that the tumor may be less sensitive to radiation owing to poor oxygenation. Given the potential advantages and disadvantages of both approaches, the roles of preoperative and postoperative radiation therapy are being actively evaluated and compared for many cancer types.[6]

The effectiveness of adjuvant radiation in soft tissue sarcomas of extremities has been clearly shown through three prospective randomized trials that have compared surgery alone with surgery and radiation. Preoperative or postoperative choice of external-beam irradiationis still a question for soft tissue sarcomas since there is no supportive data. Some of the potential advantages of preoperative external-beam radiation therapy include decreased intraoperative seeding of tumor cells, and tumor shrinkage that might facilitate later surgery. Although recurrence was expectedin this site due to poor radio-response rate of the leiomyosarcoma and positive surgical margins, relapse also occurred outside the radiation field where the surgical margins were negative. This evidence emphasizes the role of adjuvant radiotherapy in reducing the risk of recurrence in surgically treated head and neck leiomyosarcoma even without tumor positive margins.[7] The combination of wide surgical resection and postoperative adjuvant external-beam radiation therapy may be the most effective means of treatment for metastatic leiomyosarcoma.[8] Due to this reason, radiation therapy may be necessary after surgery of head and neck sarcomas.[9] While rare, leiomyosarcoma of the vertebral column must be kept in the differential diagnosis of a craniocervical junction tumor and requires a multidisciplinary treatment approach to optimize outcomes.[10,11]

**Conclusion**

Leiomyosarcoma of the head and neck are rare tumors. They tend to be intermediate or high grade and aggressive. Anticipate the possible presence of this disease and make a prudent diagnosis before the patient's life is beyond survival as early diagnosis and aggressive initial management remains the mainstay of treatment. Surgery decompression seems to reduce pain symptomatology and prevent from worsening of myelopathy.. Further studies for early diagnosis and evaluation will improve the future management and survival of leiomyosarcoma in the head and neck.

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