

Sinonasal Leiomyosarcoma: Case Report and Review of Literature

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Abstract: *Leiomyosarcoma is a malignant smooth-muscle tumour that has a predilection for the gastrointestinal and female genital tract. Sinonasal Leiomyosarcoma is exceptionally rare with no more than 100 cases reported in the English literature. It is locally fast-spreading and highly aggressive, and the prognosis is poor.*

We report a rare case of leiomyosarcoma of the nasal cavity in a 74-year-old woman who presented a swelling at the root of the nose. The histopathological and immunohistochemical studies confirmed that it was leiomyosarcoma. The patient underwent a radical surgical resection and adjuvant external radiotherapy. She is currently well and under regular follow up for the past two years with no sign of recurrence.

Radical surgery and adjuvant treatment offer the best chance for cure, nonetheless 5-years recurrence rate remains high. Sinonasal Leiomyosarcoma is a very uncommon malignant disease, characterized by high local aggressiveness. Since the incidence of this type of neoplasm is low, all patients should be treated by multidisciplinary team at referral centers.

Keywords: *Leiomyosarcoma; soft tissue sarcoma; Sinonasal; Head and neck; Radiotherapy*

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I. Introduction

Leiomyosarcoma is an unusual soft tissue sarcoma mostly affecting the uterus, gastrointestinal tract and retroperitoneum. Head and neck involvement is rare¹. Sinonasal tract Leiomyosarcoma are exceptionally rare with no more than 100 cases reported in the English literature, with most as single case reports². The origin of these types of sarcoma may be smooth muscle cells located in the skin tissue such as the piloerector muscles, or in the wall of blood vessels¹. Clinically, these tumors are very aggressive, and the prognosis is poor³. We report a rare case of leiomyosarcoma of the nasal cavity.

II. Case Report

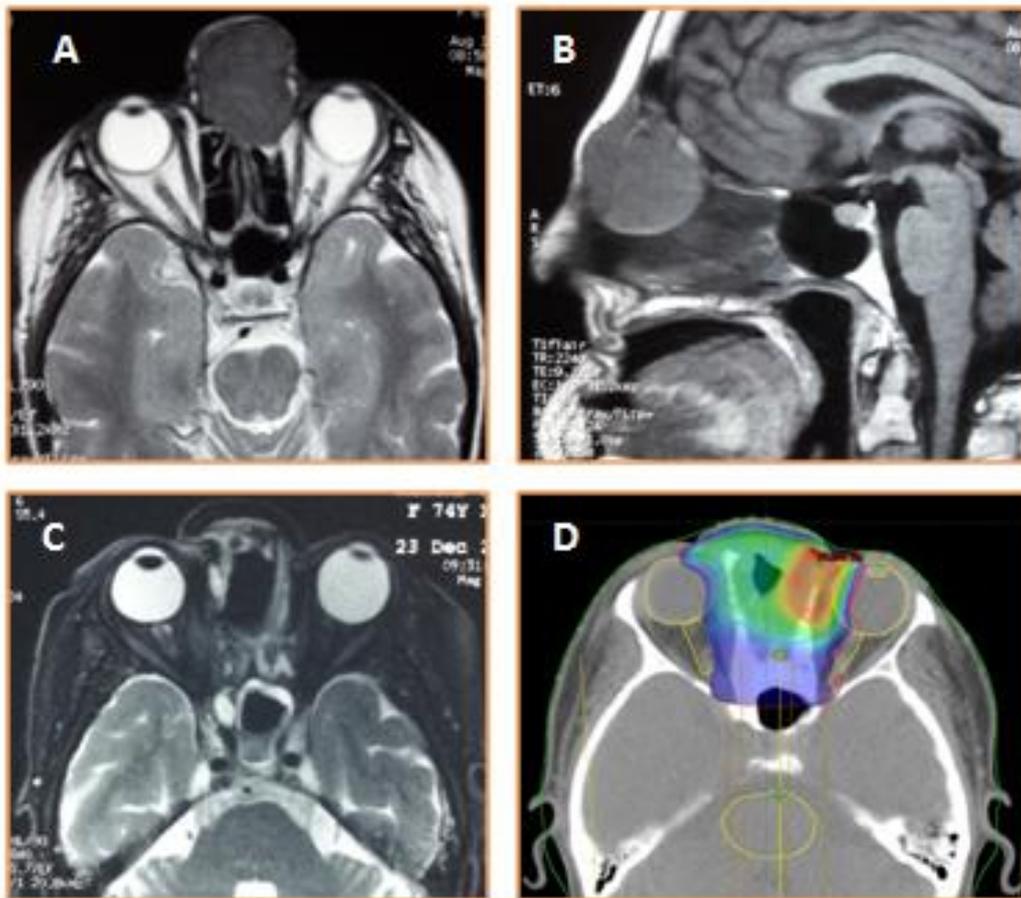
A 74-year-old woman was admitted to the Department of otorhinolaryngology with swelling at the root of the nose that had been present for seven months, accompanied by nasal obstruction but without epistaxis.

The patient had no medical history or familiar malignancy history. Clinical examination revealed a firm and painless mass at the root of the nose with conjunctivitis of the left eye. Clinical examination of the neck revealed no enlarged neck nodes.

On anterior rhinoscopy, a nonulcerated mass could be seen at the roof of the left nasal cavity. Findings on postnasal examination were normal.

Magnetic resonance imaging (MRI) with and without contrast using axial, sagittal and coronal on T1-weighted images (T1WI), T2WI revealed a contrast-enhancing tumor measuring 37 x 26 mm in diameter, involving the left nasal cavity, exceeding the nasal septum to the right with subtotal erosion of the bones own nose, filling the frontal sinus and ethmoidal cells (Figure 1A-B).

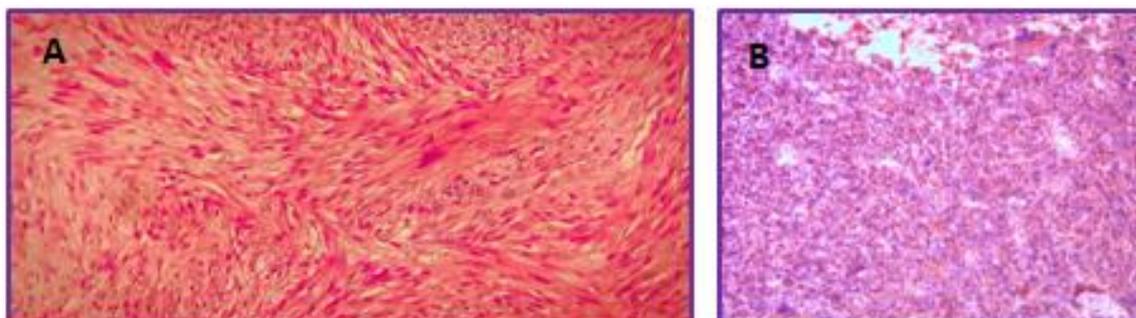
Fig 1: (A-B) Magnetic resonance imaging (MRI): tumor involving the nasal cavity, the frontal sinus and ethmoidal cells, measuring 37 x 26 mm in diameter. (C) Postoperative MRI showed no residual disease. (D) treatment by volumetric modulated arc radiation therapy: the use of two arcs allowed the optimal coverage of the target volume (red) by isodoseline 95 % (blue) without exceeding the permissible level dose in the organ at risk.



There was no evidence of regional or distant metastasis. Thoracic and abdominal CT were normal.

A transnasal biopsy revealed the diagnosis of leiomyosarcoma: The analyzed sections showed a fasciculate architecture tumor proliferation. The tumor cells have elongated nuclei, anisokaryosis and moderate hyperchromasia. The cytoplasm was eosinophilic (Figure 2 A). In places, we noted the presence of pleomorphic cells. Immunohistochemistry was positive for smooth muscle actin (Figure 2 B) and h-caldesmon. S100 protein and desmin were negative.

Fig2: (A) Malignant tumoral proliferation of sarcomatous appearance (Hex10). Immunohistochemical staining: (B) Diffuse and intense expression of smooth muscle actin (x40).



The patient underwent a radical surgical resection; the defect was reconstructed with a scalp myocutaneous flap.

Histology showed high-grade leiomyosarcoma with clear margins except posterior microscopically positive margins, Postoperative MRI showed no residual disease (Figure 1 C).

Following surgery, the patient was given an external radiotherapy delivered using volumetric modulated arc radiation therapy VMAT technique (rapidarc). The planning target volume high risk, intermediate risk, low risk received 69.96 Gy at 2.12 Gy/fraction, 59.4 Gy at 1.80 Gy/fraction, and 54 Gy at 1.64 Gy/fraction in 33 fractions respectively. Patients were irradiated once a day, five times a week. (Fig. 1 D)

The patient is currently well and under regular follow up for the past two years. Repeated CT scan and nasal endoscopy on post operative follow up revealed normal mucosa with no sign of recurrence.

III. Discussion

Leiomyosarcoma is a malignant mesenchymal tumor that derives from the smooth muscle lineage. The underlying genetic mechanisms remain unclear, and complex and unbalanced karyotypic defects are the only shared features observed across the different leiomyosarcoma subtypes⁴. leiomyosarcoma of the head and neck arise in the oral cavity, superficial soft tissues like scalp, paranasal sinuses and jaws⁵. Out of total 41 cases reported, 28 patients were males and 13 were females. The average age was 45 years. Youngest reported case is a 1 year old and the oldest 88 years⁶.

Presenting signs and symptoms are nonspecific and usually referable to the location where they arise⁴. Grossly, the lesions are smooth, firm and discretely circumscribed. Larger lesions may show focal areas of hemorrhage and necrosis. Based on a large review of leiomyosarcoma of the superficial soft tissues, the lesions of soft tissue origin 2.5 cm or larger are more likely malignant⁷.

The typical histologic pattern of leiomyosarcomas of any origin is that of intersecting, sharply marginated fascicles of spindle cells with abundant eosinophilic cytoplasm and elongated and hyperchromatic nuclei. Large leiomyosarcomas frequently contain coagulative tumor necrosis regions. Focal pleomorphism is common, and some cases show extensive pleomorphism, resembling any undifferentiated soft tissue sarcoma⁸. Most leiomyosarcomas are reactive for alpha smooth muscle actin, desmin, and h-caldesmon on immunohistochemistry⁴.

There are no randomized trials for treatment of head and neck soft tissue sarcomas. Surgical resection is the principal treatment method for the soft tissue sarcoma. 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. Neck dissection is usually not essential because of rarity of metastatic lymph node⁴.

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 radiation¹⁰⁻¹². Adjuvant radiotherapy was delivered to the base of the skull where the tumor was microscopically positive. Although recurrence was expected in 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. Due to this reason, radiation therapy may be necessary after surgery of head and neck sarcomas.

Prognosis appears to be related to the site and extent of the primary tumor. Lesions arising from the skin, nasal cavity, and larynx are associated with a better prognosis than lesions in other sites in the head and neck, probably because these sites are more amenable to complete surgical resection. Christian T. Ulrich and al, on the basis of a review of all reported cases, found, that the overall survival rate at a mean follow-up of 38.24 month was 66%. The minimal overall survival rates at 5 and 10 years were 20% and 6%, respectively¹³.

IV. Conclusion

Sinonasal Leiomyosarcoma are exceptionally rare tumors. They tend to be intermediate or high grade and aggressive. It is vital that the clinician be familiar with this lesion, anticipate the possible presence of this disease, as early diagnosis and aggressive initial management remains the mainstay of treatment. Further studies for early diagnosis and evaluation will improve the future management and survival of Sinonasal Leiomyosarcoma.

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